

MODERN TRENDS IN
DISEASES OF THE VERTEBRAL COLUMN

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MODERN TRENDS
IN
DISEASES OF THE
VERTEBRAL COLUMN

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CONTENTS

<i>Preface</i>		<i>ix</i>
<i>Chapter</i>		<i>Page</i>
1	ANATOMY AND DEVELOPMENT R WALMSLEY MD, FRCS (Ed) FRSE	1
2	CONGENITAL ANOMALIES R I HARRIS MC MB, FRCS (Cin) Hon FRCS (Eng) Hon FRACS Hon FRCS (Ed)	29
3	STRUCTURAL SCOLIOSIS J I P JAMES MS, FRCS	67
4	DEGENERATIVE DISEASES DOUGLAS H COLLINS, OBE, MD FRCP	101
5	OSTEOPOROSIS REGINALD NASSIM BM, FRCP	125
6	TUBERCULOUS DISEASE OF THE SPINE J A CHOLMELEY MB FRCS	137
7	NON TUBERCULOUS INFECTIONS OF THE SPINE J A MANTLE Ch M FRCS FRCS (Ed)	142
8	ANKYLOSING SPONDYLITIS F DUDLEY HART MD FRCP	155
9	CERVICAL SPONDYLOSIS VALENTINE LOGUE FRCS MRCP	174
10	TUMOURS OF THE VERTEBRAL COLUMN H A SISSONS MB	192
11	RADIOLOGY L S CARSTAIRS MB BS (Lond) MRCS LRCP DMR (Lond)	210
12	MANAGEMENT OF SPINAL CORD INJURIES L GUTTMANN OBE, MD MRCP	245
13	LOW BACK PAIN P H NEWMAN DSO MC FRCS	263
14	FUNCTIONAL BACKACHE WILLIAM TIGNER BM FRCP	281
15	AN ASSESSMENT OF SPINAL BIOPSY NORMAN L HIGINBOTHAM MD	287

INDEX

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PREFACE

THIS BOOK as its title implies is written on an anatomical basis, this brings many advantages because the body of information that the chapters contain can otherwise be gained only by resort to many sources. Consequently the authors are drawn from widely different fields though as might be expected most are orthopaedists.

The skeleton can no longer be regarded simply as an important supporting structure nor the vertebral column as a specialized part of this enclosing the spinal cord. Changes in the vertebral column and symptoms referable to it may, and do give the earliest indications of neoplastic, endocrinological and inflammatory diseases.

For these reasons this volume will we hope, prove useful to a wide circle of the medical profession.

We would like to take this opportunity of thanking those who have made its publication possible.

REGINALD NASSIM
H JACKSON BURROWS

October 1958

CHAPTER 1

ANATOMY AND DEVELOPMENT

R WALMSLEY

INTRODUCTION

THE GENERAL features of the adult vertebral column as a whole and the composition and structure of its component parts are so well known that many of them are considered to be outside the scope of this chapter

Man has developed the orthograde position to its most perfect stage and during the course of evolution the vertebral column has shown extensive alterations from the basic form present in pronogrades. Among bipeds man is unique because he alone is capable of fully extending the thighs on the trunk in normal posture and adopting an easily maintained stance which allows him to use his upper limbs freely and independently the anthropoid apes (also orthogrades) are unable to do this and they use their upper limbs not only in locomotion but also in standing. In mammals generally the average number of presacral segments is 27 whereas in man the number is reduced to 24 in about 95 per cent of individuals. As part of the reduction (two vertebrae) occurs in the lumbar region it allows orthogrades to maintain the erect posture more easily and provides for a closer approximation between the thoracic cage and pelvis with a corresponding reduction in the length of the antero lateral abdominal wall.

The bodies of the vertebrae have been considered as forming four pyramids when the column is viewed from the front as the width of the bodies is stated to increase along the cervical column from axis to the seventh cervical vertebra then decrease to the fourth thoracic from where there is a gradual increase in width to the fifth lumbar vertebra before coming to the triangular shaped sacrum and coccyx. The narrowing in the upper thoracic region is not particularly obvious although the antero posterior depth may be seen to increase down to the second lumbar below which there is a slight diminution in the measurement. Kohler (1935) has observed that the body of every vertebra except the last lumbar is taller than that which lies immediately above it.

Each region shows its own specialized vertebral form and the intervertebral joints show a range of types that vary from the synovial joints between the atlas and the axis to the synostosis of the vertebrae in the sacrum and the coccyx. This regional difference is apparent to some extent in all higher vertebrates and may well be regarded as an expression of function of the region.

Curvatures of the spine

The antero posterior curves in the newborn child and an adult man are shown in Fig 1. In the newborn child the sacrum is obviously more in alignment with the upper part of the column than in the adult and this feature contributes to the small size of the pelvis in young children and the consequent intra abdominal position of the bladder. The inclination of the sacrum shows great

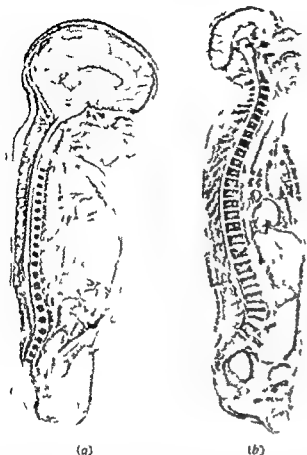


FIG 1—(a) Median section of new born child. The 2 primary curvatures are present but their concavities are not prominent. The upper curvature extends from the atlas to the lower border of the fifth lumbar vertebra and the lower embodies the sacrum and the cartilaginous coccyx. The angulation of the sacrum is about that commonly seen in newborn children. (b) Median section of adult man presents a mild secondary cervical curvature and a more prominent lumbar curvature. The angulation of the sacrum is within the normal range. In this specimen the spinal cord extended to the lower border of the second lumbar vertebra, which is marked by a piece of tapered tubing. A large Schmorl's node is present in the upper part of the first lumbar vertebra. (Fig 1b is reproduced by courtesy of the Editor *Edinb med J*)

variation in different adults and different authors measure the lumbosacral angle in different ways. It is sometimes defined as the angle between the long axis of the lumbar spine and the sacrum and sometimes as the angle between the plane of the upper surface of the sacrum and a horizontal plane. Whichever method is used the variations that may occur in the range of inclination of the sacrum would appear to be in the region of 50 degrees. Secondary (or compensatory) curves develop in the cervical and lumbar regions with the raising of the head and with the extension of the lower limbs when a child begins to walk—and it is noticeable that the cervical curvature is not so pronounced as the lumbar. Both these curves are dependent on the posture of the body because as the cervical curvature is lost when the head is flexed so too the lumbar part of the column becomes flattened and more in line with the sacrum when the subject sits upright in an ordinary straight chair or when the trunk is flexed on the thighs as in touching the ground with the finger tips. More important from the clinical diagnostic point of view is the change in the lumbar curve when the extended leg (or straight leg) is raised from the supine position (Charnley 1951, Armstrong 1952). The greater part of the movement of straightening the lumbar column occurs at the last interlumbar and the lumbosacral joints and the corresponding intervertebral discs have an especial tendency to degenerative changes. Capener (1944) and Keegan (1953) among others have drawn attention to the action of the hamstring muscles in flattening the lumbar curves in sitting. Similarly Keegan believes that the psoas major muscles through their attachments to the front of

each lumbar vertebra and the first sacral vertebra in the upright position. The secondary curve is a result of the position of the intervertebral discs, but the first of the fifth lumbar vertebra is deeper than the back and the same holds true for the first and third lumbar vertebrae (Fig. 11).

The series of alternate vertebrae and discs are joined together to form an intricately constructed spinal column that not only supports the trunk but is also endowed with flexibility and resistance. Each component, whether it be bone, intervertebral disc, synovial joint or ligament, plays a particular role in endowing the column with the characteristic of an organ and one component will reflect itself in an up or down of the function of the whole.

The lateral curvature often present in the mid thoracic region is a common convex to the right in right handed people and the reverse in left handed people. This curve has also been associated with the position of the thoracic aorta lying on the left of the intermediate thoracic vertebrae. If however the curvature is deduced from the direction of the spinous processes and if a lateral rotation of the vertebrae is assumed, then the truth of the statement quoted by Kilshaw and Ollerenshaw (1954) is to be remembered, namely that the thoracic vertebral bodies rotate away from the concavity and the spinous processes towards it.

Variations in vertebrae

Although all vertebrae have the same basic form initially they soon assume features that are characteristic of the body segment to which they belong. Thus in about 90 per cent of people there are 7 cervical, 12 thoracic, 5 lumbar, 5 sacral and 4 coccygeal vertebrae but in the remaining 10 per cent a vertebra assumes partially or completely the characters of an adjacent vertebra. It is in the transitional regions where the most obvious changes tend to occur. Although for example the twenty fifth vertebra forms the first sacral in 95 per cent of people the twenty sixth does so in 3 per cent and the twenty fourth vertebra in 1 per cent. Bardeen (1905) has described how the vertebra that is to form the first sacral irrespective of whether it is the twenty fourth, twenty fifth or twenty sixth in the vertebral series takes on the characteristic form of this vertebra in an early stage. In the thoraco lumbar region the twentieth vertebra which normally forms the first lumbar bears rudimentary ribs in about 6 per cent of people and the eighteenth vertebra in about 2 per cent of people is devoid of ribs. The variations in the development of the costal element of the seventh cervical vertebra are well recognized and may be of clinical importance when as in about 1 per cent of people it forms a definite cervical rib which is crossed by the lower trunk of the brachial plexus (Le Double 1912 and others).

EARLY DEVELOPMENT

The developmental history of the vertebral column is complex and the vertebrae and the discs cannot be dissociated from each other. In their structure and function the discs and vertebrae likewise must be recognized as two elements of one unit and it certainly appears to the writer that there can be no logical consideration of the one either in health or disease without a simultaneous consideration of the other.

When the embryo is about 5 weeks old there lie along each side of the neural tube and notochord the prominent series of somites (Fig 2) The somites originate by segmentation of a continuous column of mesoderm termed the paraxial bar and within each somite shortly after its formation there appears a small cavity termed the myocoele While the myocoele is present the somite has a triangular form with medial lateral and ventral walls (Fig 3)

It is the cells of the lower part of the medial wall of the somite that are concerned with the development of the axial skeleton and consequently this part of the somite bears the name of sclerotome (Goodsir, 1857) The remainder of the



FIG 2—A photograph of a model (Ziegler) of a human embryo approximately 7 millimetres long The somites are seen as a series of prominent cubic elevations along the dorsal wall (Reproduced by courtesy of the Editor *Edinb med J*)



FIG 3—Schematic section of embryo On the left side of the diagram a somite is shown with intact sclerotomic and myotomic parts On the right side the sclerotomic cells are shown migrating medially both ventral and dorsal to the neural tube (Reproduced by courtesy of the Editor *Edinb med J*)

somite is termed the myotome or dermomyotome The cells of the sclerotome migrate medially and passing both dorsal and ventral to the notochord meet those of the opposite side in the mid line and so form a continuous membranous mesodermal column around the notochord (Fig 3) some of the cells migrate to the sides of the neural tubes where they form the vertebral arch Remak (1855) postulated resegmentation of the developing vertebral column and described how each sclerotome contributes to the formation of parts of two vertebrae and one intervertebral disc Von Ebner (1888) noted that each sclerotome early in its development is divided into cranial and caudal halves by a partial

EARLY DEVELOPMENT

transverse fissure which may appropriately be termed the sclerotomic fissure (Fig 4). The sclerotomic cells that bound this on its cranial and caudal sides proliferate to form a well defined dense band that is the forerunner of the disc and may be termed the *primitive intervertebral disc*. The caudal part of one sclerotome then unites with the cranial part of the adjacent sclerotome to form a primary vertebra. In consequence of this revised segmentation the vertebrae alternate in position with the myotomes. This mode of formation of the primitive vertebrae and intervertebral discs has important basic functional implications as the muscles that arise from one myotome are thus related to the parts of two skeletal elements and an intervertebral disc on which they act.

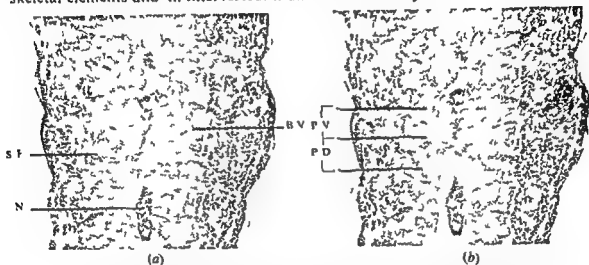


FIG 4—Coronal section of lumbosacral region of 8 millimetre human embryo. The notochord (N) is undulant and therefore not cut throughout its length. The blood vessels (BV) are segmental and therefore related to the primitive vertebra (PV) which is formed by adjacent parts of two sclerotomes. The primitive disc (PD) is seen as a dark band and within it is the sclerotomic fissure (SF). Note the relative sizes of the primitive vertebrae and the discs at this stage ($\times 40$). (Reproduced by courtesy of the Editor *Edinb med J*)

It may be postulated further that the adjacent parts of any two vertebrae and the intervening intervertebral disc constitute along with the muscles that arise from the same somite not only a developmental unit but also a functional unit (Fig 5).

The sclerotomic fissure within the primitive disc and opposite the middle of the myotome is shown in Fig 4. It is transient and all trace is lost soon after the 8 millimetre stage. The anlage of the body of the vertebra formed from two sclerotomes is represented at an early stage by a mass of loosely arranged cells. The primitive body and the primitive disc are of about equal length (Fig 4). The size of a structure at this stage gives no indication whatsoever of its definitive size.

The notochord both in phylogeny and ontogeny is the forerunner of the vertebral column and though all evidence of the notochord is normally lost within the vertebral bodies it is intimately concerned with the formation of the intervertebral discs. The intervertebral disc has the most complex development of all the joints largely because of the notochord within the primitive disc. In the 15 millimetre human embryo the notochord extends from the clivus of the

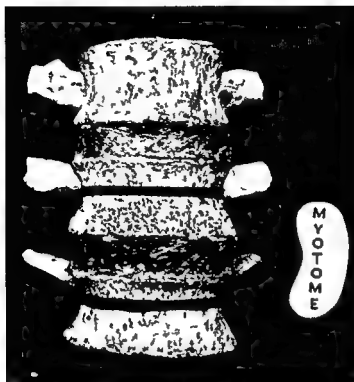


FIG 5—Photograph of the lower 3 lumbar vertebrae with the intervening intervertebral discs. The parts of the adjacent vertebrae which have been developed from the corresponding sclerotomes of the right and left sides lie between the black lines and are stippled. The position of a myotome of the same segment is indicated (*Reproduced by courtesy of the Editor Edinb med J*)

skull to the surface of the coccyx. In the skull it passes obliquely through the basi occiput (Fig 6a) emerges on the pharyngeal surface of the skull and re enters the sphenoid to terminate in the region of the dorsum sellae. The peripheral part of the primitive disc forms the annulus fibrosus whereas the central part with the notochord forms the nucleus pulposus. Within the peripheral region of fibroblasts there is an *inner zone* which although relatively small at this stage will by the proliferation of its cells, participate most actively in the formation and growth of annulus and nucleus pulposus (Prader 1947b Peacock 1951 1952 Walmsley 1953). The outer fibrous zone of the disc is clearly demarcated from the inner zone which assumes an appearance rather like pre cartilage and has been termed hyaline cartilage by Prader (1947b) and specialized embryonic cartilage by Peacock (1951). Prader's concept of a continuous cartilaginous column at this stage appears to be open to question.

Development and ossification of a vertebra

The body of a vertebra as already stated is formed by the union of mesenchyme from parts of adjacent cranial and caudal sclerotomes so that the primitive vertebrae alternate with the myotomes or muscle rudiments. When the embryo is about 12 millimetres long the mesenchyme of the vertebral body differentiates as cartilage from 2 chondrification centres 1 on each side of the notochord which soon fuse (Bryce 1915). Chondrification of each side of the vertebral arch

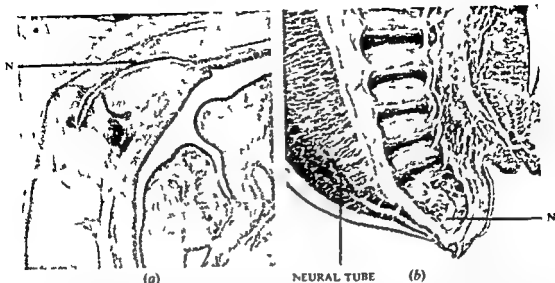


FIG 6—Sagittal sections of 15 millimetre human embryo (a) Showing the notochord (N) passing from the odontoid process into the cranial cavity and then passing obliquely through the basi occiput to continue on the pharyngeal surface of the skull (b) Showing notochordal cells (N) reaching the dorsal surface of the coccyx

begins in a centre near the junction of pedicle and lamina. Each arch centre soon fuses with the body in the formation of a cartilaginous pedicle but chondrification extends backwards more slowly and the vertebral arch remains open dorsally until the fourth month of intra uterine life when the laminae meet and fuse. As a cartilaginous spinous process forms and grows backwards from the junction of the bilateral laminae, so too does a transverse process grow laterally from the junction of pedicle and lamina.

Ossification in cartilaginous bone conforms to one of three distinct patterns. First as in an epiphysis an ossific centre appears within the cartilage, and enchondral ossification gradually extends towards the surface. Secondly diaphyseal bone as it may be termed, is typically seen in the diaphysis of a long bone and there the *first* bone is deposited on the surface of the cartilaginous shaft after it has become calcified. Thirdly some bones appear to combine these two methods of ossification either synchronously or one before the other.

Reporting on a series of 3 and 4 months old foetuses Mutch and Walmsley (1956) concluded that the ossific centre that appears in each half of the vertebral (neural) arch is *always* on the inner surface of the arch that is, bone of the diaphyseal type. The centre for the centrum usually appears on the posterior surface of the body of a vertebra and only occasionally additional ossific centre or centres appear simultaneously within its substance. This unorthodox concept of vertebral ossification must be appreciated for a proper understanding of some pathological conditions of the vertebral column.

The postero lateral parts of each body are ossified from the vertebral arches separately from the intermediate part or centrum (Fig 7). Fusion of the bone of the neural arches with the centra and obliteration of the neuro central joints begins in the lower cervical vertebrae at about 3 years and the process, extending upwards and downwards is completed in all regions by about the sixth or seventh year.

Ossification of the centrum

In lumbar vertebrae of 3 months old foetuses (Fig 8a and b) the cartilage in the intermediate part of the centrum becomes calcified and the calcification reaches the posterior surface before the anterior surface. In agreement with Streeter (1949) that such calcified tissue stimulates the formation of periosteal bone the first site of bone in a vertebral body is usually its *posterior surface* in relation to the calcified cartilage. Vascular canals are however, present within the cartilage before ossification and in this feature vertebral ossification differs from diaphyseal ossification. Coincident with the formation of the periosteal bone the calcified cartilage is excavated and large spaces termed secondary areolae appear within

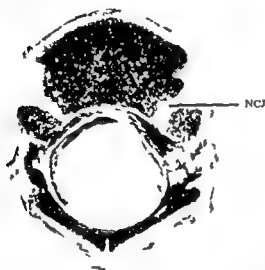


FIG 7—Lumbar vertebra of a young child divided to show the neuro central joint (NCJ). The laminae which in the lumbar region fuse soon after birth are seen to be fused on their deep aspects

A secondary breakdown of the periosteal collar of bone allows the calcified cartilaginous mass to be invaded by additional vessels such a stage is represented in a lumbar vertebra of a 4 month old foetus (Fig 9a and b) on the cartilaginous walls of the large secondary areolae bone is deposited. When cartilaginous calcification of the cartilage reaches the front of the cartilaginous vertebral body bone begins to be formed on this surface (Fig 9a). The periosteal bone both on the front and the back of a vertebra shows an irregularity of form often with spur like processes (Fig 10a and b) characteristic of diaphyseal long bone formation.

The massive excavation of the cartilage within the vertebra typical of diaphyseal ossification would seem inconsistent with the ossification from one two or more separate endochondral centres. The notochordal mucoid streak persists for some time within the cartilage of the vertebral body and delays the normal sequence of changes in cartilage and the formation of bone.

From observations in Scheuermann's disease Knutsson (1948) concluded that the antero posterior growth of a vertebra occurs mainly by a deposition of bone on its anterior surface this may be correlated with thickening of the front of the annulus fibrosus of the lumbar discs with age and the consequent apparent backward displacement of the nucleus pulposus.

The upper and lower surfaces of the ossified part of a young vertebra have a series of ridges and furrows which are covered in life with cartilage. Within the

EARLY DEVELOPMENT



FIG 8—Sagittal section of lumbar vertebra of 94 millimetre human foetus (a) ($\times 20$) Showing the eccentric position of the zone of calcification and (b) ($\times 40$) the thin lamina of bone on the posterior notched surface (Reproduced by courtesy of the Editor *Edinb med J*)

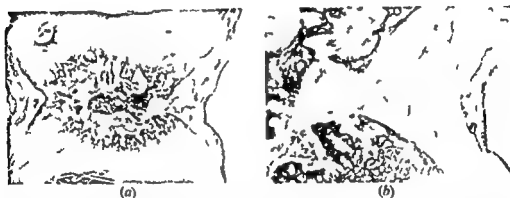


FIG 9—Sagittal section of lumbar vertebra of 144 millimetre human foetus (a) ($\times 12$) Showing the eccentric position of the bone which has just begun to be formed on the anterior surface and (b) ($\times 35$) bone on the posterior surface fenestrated and transmitting vessels (Reproduced by courtesy of the Editor *Edinb med J*)

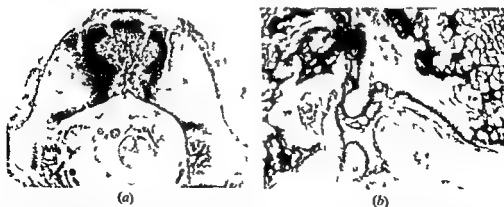


FIG 10—Transverse section of the lumbar vertebra of 124 millimetre human foetus. A thin lamina of bone is present on the posterior surface and is shown at higher magnification in (b) ($\times 55$). The mushroom shaped form of the bone in the vertebra is apparent in (a) ($\times 8$) (Reproduced by courtesy of the Editor *Edinb med J*)

peripheral part of the cartilage related to each furrow there appears a separate secondary centre of ossification and these centres later fuse and form the annular epiphyses of the vertebral body. Beadle (1931) recorded Schmorl's observations that the secondary centres may appear as early as the eighth year and the fusion of the epiphyseal ring to the diapophysis of the vertebra is usually complete at the twenty fifth year and sometimes several years earlier. Bick (1952) took exception to the use of the term 'epiphyses' for these annular rings but in view of comparative studies it is considered that this is an erroneous conception of them.

It is well recognized that in most pronograde mammals the epiphyses of the vertebral bodies are represented by complete plates. The ossification of each plate is from multiple centres which appear in the dorsal part of the periphery and extend ventrally. The human epiphyses may therefore be regarded as a modified form of mammalian epiphysis in which the ossific process failing to invade the central zone of cartilage allows the epiphysis to assume its peripheral annular form. Its limitation to the rim of the vertebral body ensures that the entire vertebra may be associated with the nutrition of the intervertebral disc throughout life and is not dependent between the eighth and twenty fifth year on the meagre circulation in thin plate like epiphyses (Walmsley 1953).

The centres for the centra appear first in the lower thoracic region at about the tenth week the process spreads upwards and downwards until by about the thirtieth week centres have appeared in all bodies except the coccyx which is not ossified until after birth (Fig 1a).

Ossification of the vertebral arch

There is one primary centre of ossification for each half of the vertebral arch. These centres first appear in the upper cervical region during the seventh week of intra uterine life and having a cranio-caudal sequence are apparent in the sacrum not less than 3 months later. During early development there may therefore be a considerable difference in the stage of ossification even of adjacent vertebral arches (Fig 11).

Before ossification begins the vertebral arch is avascular, thus differing from the body of the vertebra which shows numerous vascular canals already referred to which are seen as clear spaces (Fig 11). The first indication of ossification is localized calcification in the part of the arch that lies between the superior and inferior articular processes known as the *pars interarticularis*. When calcification reaches the nearby antero medial surface of the *pars interarticularis* subperiosteal bone is laid down and thereafter the calcified cartilage is invaded by blood vessels and partly excavated. Calcification of the matrix extends in all directions from its initial focus upwards and downwards into the roots of the articular processes forwards into the pedicle and backwards and laterally towards the posterior and lateral aspects of the *pars interarticularis* and gradually reaches the perichondrium in these regions. Wherever it does so the perichondrium assumes an osteogenetic function and deposits subperiosteal bone (Fig 11). The sequence may be seen in some measure at least in a single sagittal section of the lumbar vertebrae of a 101 millimetre foetus (Fig 11). While the deposition of subperiosteal bone continues there is massive excavation of the central cartilage of the *pars interarticularis* (Fig 11b) but upon the small islets of calcified cartilage that remain the primary medullary bone of the trabeculae is laid down.

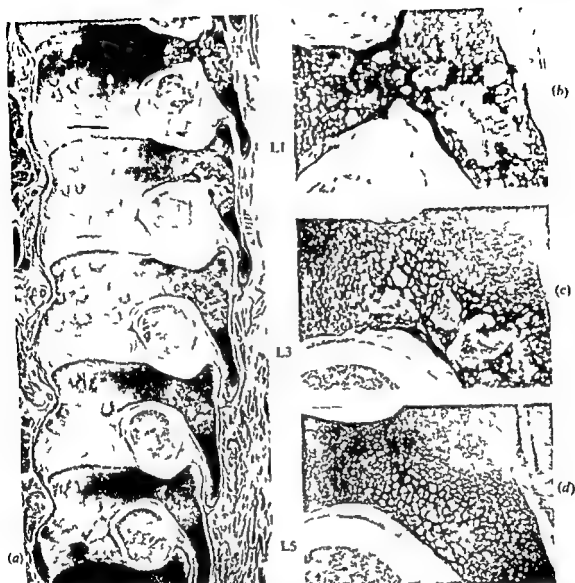


FIG 11 —(a) Sagittal section of a lumbar vertebra of 101 millimetre human foetus ($\times 7$). The section passes through the pars interarticularis of each vertebra and the superior and inferior articular processes are clearly seen. Ossification is just beginning in L 5 (d) and is most advanced in L 1 (b). The higher power sections of L 5, 3 and 1 show the ossific changes more clearly ($\times 30$). (Reproduced by courtesy of the Editor *The Lancet*.)

Reconstructions of lumbar vertebrae of three foetuses (75, 93 and 101 millimetres long) have shown that the calcified cartilage along with the first formed bone in the pars interarticularis together constitute a hemispherical mass (Mutch and Walmsley 1956) unlike the hour glass form that Willis (1931) described as erroneously suggesting two separate centres.

Sections through the pars interarticularis of older foetal and postnatal vertebrae show great variations in the thickness of the cortical bone and in the density of the medullary trabeculae in specimens of about the same age.

These observations are in striking contrast to the statements of Hitchcock (1940) who stated that ossification of the vertebral arch is preceded as in all endochondral ossification elsewhere by the invasion of vessels from the laminar

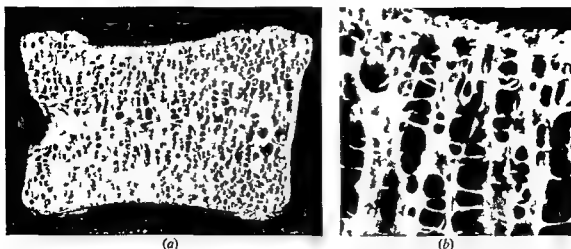


FIG 12—Sagittal section of lumbar vertebra (a) Showing the entire surface of the body with its posterior surface towards the left. A line passes through a venous channel (b) Showing a higher power ($\times 16$) of the upper part of the vertebra (Reproduced by courtesy of the Editor *Edinb med J*)

arteries and the periphery of the isthmus (interarticular) region remains cartilaginous up to and after birth

As long as Bardeen's (1910) statement is perpetuated that ossification in the vertebral arch is endochondral and his figure showing this ossification within the substance of the cartilage of the arch continues to be reproduced so too will there be misapprehension about possible anomalies in the neural arch. Bardeen's statement that sometimes in the fifth lumbar vertebra there are two centres for the arch (for which he gives no authority) is inconsistent with the pattern of ossification involved. This pattern is basically opposed to the appearance of either single or double primary endochondral ossific centres within the vertebral arch which explains the failure of Rowe and Roche (1953) to discover a single instance of cleft vertebral arch in over 500 stillborn and neonatal cadavers. If a cleft vertebral arch has a congenital factor it is due not to two centres of ossification but possibly to a congenital weakness in the bone of the pars interarticularis.

Fusion of the bony laminae begins in the lumbar region soon after birth and spreads slowly upwards and more slowly downwards, being completed in the cervical region by the second year and in the sacrum (above the hiatus) by about the tenth year.

Besides secondary ossific centres for the annular epiphyses of the body there are secondary centres for the spinous, the transverse and the mamillary processes (of lumbar vertebrae) which appear about the fifteenth year and fuse about the twenty-fifth year.

The manner in which a vertebra ossifies is considered to be closely correlated with the growth of the vertebral canal. The bone that is first formed in arches and centrum is rapidly absorbed and it is only after the ossific centres have appeared that a gap appears between the developing spinal cord and the arch and a potential space is created.

INTERNAL STRUCTURE OF VERTEBRAE

The architecture of the body of a vertebra accords with the compression stresses it has to withstand. The trabeculae are arranged parallel to one another and

in line with the long axis of the column, and binding them together are finer transverse struts (Fig 12). The porous nature of the upper and lower surfaces of the bodies is correlated with the nutrition of the intervertebral disc (p 10). These surfaces except where related to the bone formed by the annular epiphyses are covered by plates of hyaline cartilage which in the immature bodies serve as a growth zone (Harris and Menden 1954) but are unlike normal epiphyseal cartilage as they persist after growth has ceased. The internal architecture of the vertebrae has been extensively studied by Gallois and Japrot (1925) and its histology by Bick (1952). More recently Farkas (1954) has given an excellent account of the intravertebral structure in the different regions drawing attention to the original observation by Beidle (1931) that the unfused annular epiphysis is only loosely attached to the circumference of the vertebra and can be easily separated from it.

The intervertebral disc

In the cervical and thoracic regions the nucleus is approximately central but in the lumbar region it is slightly more posterior. Its size, structure and composition vary with age. In the lumbar disc of the newborn the nucleus in antero-posterior section is at least four times as thick as the annulus behind or in front of it (Fig 17a) and in the adult where it is represented by fibro cartilage it is relatively much larger than schematic drawings often depict. It has been stressed repeatedly that the structural complexity of the intervertebral disc is in keeping with its specialized functional requirements and that not altogether surprisingly, it undergoes more obvious changes during prenatal and postnatal life than any other joint.

Annulus fibrosus

In the lumbar region the adult annulus fibrosus is formed by a series of laminae which have an intricacy of pattern that almost defies description. The most peripheral laminae are formed of pure collagenous tissue and at the front and back these fibres of the annulus blend with the strong anterior and the weaker posterior longitudinal ligaments. Within the fibrous covering of the annulus lie *fibro cartilaginous laminae* which form its major part. In a transverse section of the disc of a young adult (Fig 13) the lamination of the annulus is readily apparent and its innermost fibres blend with the nucleus pulposus tissue from which the annulus is not sharply demarcated. The laminae do not form complete rings and though they may encompass about half of the nucleus they either become attenuated and finally cease or bifurcate and enclose the termination of another lamina (Figs 13 and 15). At the sides of the annulus there are numerous strong fibrous bands connecting adjacent laminae. It is this pattern of dovetailing and anastomoses between laminae that gives the annulus its complicated pattern. The laminae become noticeably thicker as they are traced from the periphery towards the nucleus.

In the lumbar region some 12 to 16 laminae can usually be enumerated anteriorly and at the sides these are often supplemented by several deeply situated laminae having a relatively short antero-posterior course. On account of the posterior excentric position of the nucleus in adult lumbar discs the laterally placed laminae in their course backwards become increasingly slender (Fig 13) and often bifurcated. In the posterior part of the annulus they join with each other to form a narrow zone of great complexity that lies directly behind the

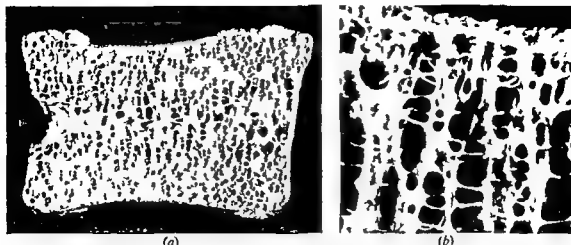


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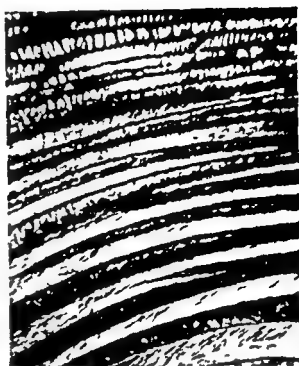


FIG. 15—Part of the annulus fibrosus of a full term foetus as seen with polarized light. This conforms to the pattern of the adult annulus ($\times 45$). (Reproduced by courtesy of the Editor *Edinb. med. J.*)

Tondury (1944) and Prader (1947) maintain that ligaments are endowed with this basic quality of self differentiation, but Ubermuth (1929) considered that the fibre system of the intervertebral discs is formed as a direct response to vertebral function and that the intricate formation of the fibro cartilage component occurs only after birth when the child begins to sit and to walk. This conception of the formation of joint ligaments in general still receives some support (Happy Macrae and Naylor, 1954) which appears to the writer completely unjustified. The basic pattern of ligaments, no matter how intricate appears to be determined early in foetal life and it is on the basic pattern that the tension, and it may be compression forces experienced during movement that exert a modifying influence.

In a 43 millimetre embryo (Fig. 16a) the fibroblasts have attained a typical elongated form but at this stage have elaborated little collagen. In a coronal section of the annulus it is apparent that the cells are orientated in such a manner that they may be classified into 2 groups: those having their long axes directed upwards and to the right and those having their long axes directed upwards and to the left. This coronal section is thick enough to include several laminae and that is why the annulus presents a lattice like appearance. This is typical of the form of annulus fibrosus in embryos of this age and the general direction of the fibroblasts resembles the arrangement of the collagenous fibres in the adult annulus. In Fig. 16b and c, photographs are included of coronal sections of the annulus of a 5½ month old foetus and a 5½ year old child respectively and their correspondence with the younger specimen is apparent. The cruciate arrangement of the fibres of laminae of the annulus to which reference has been made offers an excellent example of self differentiation possessed by ligaments.

Posterior part of annulus fibrosus

Attention has already been directed to the excentric position of the nucleus pulposus in adult lumbar intervertebral discs and to the associated relative

nucleus The eccentric position of the nucleus pulposus contributes to the relative weakness of the posterior part of the annulus

Lamination of the annulus fibrosus becomes apparent in very young embryos (15 millimetres long) and in the full term foetus (Fig 15) resembles that in the adult The intricacy of the annulus is further increased by the difference in direction of the collagenous fibres in adjacent laminae In the lumbar region the fibres of contiguous laminae cross each other obtusely and the main mass of fibres in any one lamina passes at an angle of about 30 degrees to the horizontal plane (Labalt 1835) (Fig 14)

The function of the annulus would appear to be two fold to restrict and regulate movement and to enclose and retain the nucleus pulposus The arrange

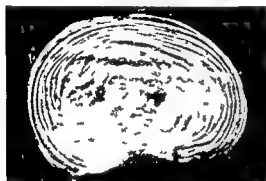


FIG 13—Transverse section of the lumbo sacral intervertebral disc of a young adult The intricate pattern of the laminae of the annulus is apparent and is in contrast to the appearance of the nucleus which is nearer the back than the front (Reproduced by courtesy of the Editor *Edinb med J*)



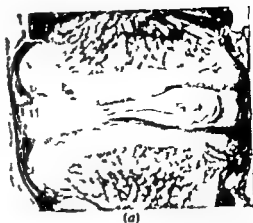
FIG 14—Photograph of a dissection of the annulus fibrosus which shows the different obliquity of the fibres in adjacent laminae A lumbar nerve is seen emerging behind the annulus on the left side (Reproduced by courtesy of the Editor *Edinb med J*)

ment of the fibres enables the annulus to control the torsional movements of the vertebral column and to assist in limiting flexion and extension in any plane The cruciate pattern accords with the arrangement of tissues in other walls for example the abdominal wall that are subjected to pressure from within and it is postulated that the criss cross arrangement of the annular laminae is to be correlated primarily with the internal pressure or thrust exerted on them by the nucleus pulposus when it is subjected to compression forces

The self-differentiation of ligaments

The self differentiative powers of bone are well recognized and all ligaments examined by the writer were endowed with similar properties Brockmann (1942)

Fig 17—Median sections of lumbar intervertebral discs of (a) full term foetus ($\times 6$) (b) 11 year old child ($\times 5$) and (c) 56 year old man ($\times 5$). The photographs are to illustrate the changes that occur in the posterior part of the annulus with the development of the lumbar curve and the relative approximation of the posterior parts of the vertebrae (Reproduced by courtesy of the Editor *Edinburgh med J*)



(a)



(b)



(c)

DEVELOPMENT OF NUCLEUS PULPOSUS

Keyes and Compere (1932) reviewed some aspects of the development of the intervertebral disc and concluded that the nucleus pulposus is initially formed by the proliferation and mucoid degeneration of the notochordal cells and that there is a subsequent invasion of fibro cartilage from the tissue which surrounds the notochord. Walmsley (1953) arrived at conclusions that in many ways resemble those of Keyes and Compere and of Peacock (1951, 1952).

The notochord of the 17 millimetre human embryo, while still of almost uniform thickness, shows localized aggregations of cells at the level of the developing discs (Fig 18) followed later by a diminution of cells in the intravertebral region. A little later still the notochordal cells in the intervertebral region begin to form localized expansions which become very evident in sagittal sections and the part of the notochord within the cartilaginous vertebra loses its cellular character and is replaced there by a mucoid streak (Fig 19).

The appearance presented by the localized aggregations of notochordal cells in the developing discs of embryos in which the mucoid streak is present suggests a passive displacement of the cells from the vertebral bodies into the intervertebral regions (Kölliker 1867 Williams 1908). Examination of serial sections has convinced the writer that the aggregations of notochordal cells in the intervertebral discs is not dependent on their displacement from the intravertebral regions

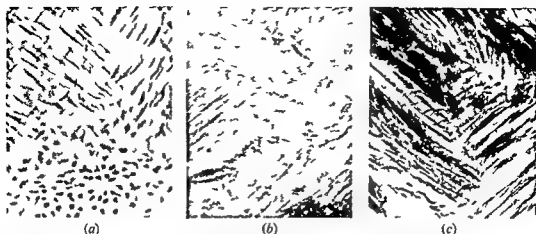


FIG 16—Three photo micrographs of coronal sections of the annulus fibrosus to illustrate the cruciate arrangement of the laminae (a) 43 millimetre embryo in which fibroblasts have an elongated form and are orientated in planes like those in older specimens. Little collagen elaborated at this stage ($\times 225$). (b) 170 millimetre foetus (5½ months) showing cruciate arrangement of collagen fibres and fibroblasts ($\times 44$). (c) 5½ year old child showing denser collagenous tissue and relatively few fibroblasts ($\times 48$). (Reproduced by courtesy of the Editor *Edinb med J*)

thinness and weakness of the posterior part of the annulus fibrosus. During the examination of microscopic sections of foetal and postnatal lumbar intervertebral discs a tissue change may often be seen in the posterior part of the annulus the consequence of the formation of the secondary lumbar curvature. Before the secondary curvatures appear the length of the fibres of the anterior and posterior parts of the annulus is approximately the same and the laminae pass with a gentle sweep between the contiguous surfaces of adjacent vertebrae (Fig 17a). The formation of the secondary lumbar curvature causes a widening of the interval between the anterior parts of the bodies of the lumbar vertebrae and an approximation of their posterior parts. To this approximation the posterior fibres of the annulus have of necessity to adapt themselves by assuming a U shaped course between vertebrae (Fig 17b).

In the central angular interval between the sharply curved fibres of the posterior part of the annulus additional fibres which have a more direct course between the vertebrae are developed the deepest of which may have a convexity directed towards the nucleus (Franceschini 1947). When the joint is extended the oblique course of the fibres in the posterior part of the annulus renders them subject to compression forces to which the fibrous tissue of the annular laminae is ill suited. Fibrous tissue is pre eminently adapted to withstand tensile forces but when unsupported by a firm matrix as in bone and cartilage may undergo degenerative changes when compressed. An expression of these changes is the rupture of the fibres at the summit of their curvature as shown in Fig 17c. The formation of the lumbar curvature necessitates a degree of adaptive change in the associated intervertebral discs that is unequalled in other regions and is most evident in the disc between the fifth lumbar vertebra and the sacrum. The posterior part of the annulus fibrosus of a lumbar disc may therefore be considered as a site of potential weakness. This regional weakness is dependent in part on the thinness of the annulus and in part on the degenerative changes that compression may induce in the collagenous fibres.

Malmgren and Sylven (1952) has a high content (30-36 per cent dry weight) of the polysaccharide, chondroitin sulphate a low protein fraction and salts in addition to the water, the polysaccharide around the fibrils endows them with their hydrophilic properties

The sequence of age changes in the nucleus pulposus is essentially an account of the maturation of collagen and its replacement of the mucoid material (Fig 21b and c) The main mass of the nucleus pulposus of the full term foetus is formed of mucoid material, into the periphery of which collagenous fibres extend from the surrounding capsule of fibro cartilage, embedded in the mucoid material are scattered notochordal cells The total disappearance of notochordal cells appears to occur before the end of the first decade, concurrently the mass of mucoid material ceases to grow and is replaced by fibro cartilage There is a differentiation of collagenous fibres *in situ* besides invasion of the nucleus by the surrounding fibro cartilage The cartilage cells of the replacing tissue are derived mostly from the surrounding annulus fibrosus but also from the cartilage plates that cover the upper and lower surfaces of the vertebrae In young people there is a sharp line of demarcation between the cartilage plate and the nucleus pulposus, but in older discs cartilage cells may be seen invading the nucleus pulposus The notochordal cells on the one hand are responsible for the elaboration of the mucoid material by their action on the surrounding tissue and on the other prevent the invasion of the mucoid material by the surrounding fibro cartilage The short life of the notochordal cells is a feature of their existence and, if they possess the qualities described their early death is of major importance in the life of man

Happey, Macrae and Naylor (1954) carried out an x ray crystallographic investigation of the changes that occur in the nucleus pulposus and annulus fibrosus with age They reported that in the nuclei of young people the fibrils have a random distribution but that with the increase in years they show a definite orientation

Erlacher (1952) has described a technique of demonstrating the zone of the nucleus by injecting 1-2 millimetres of radio opaque fluid through a lumbar puncture needle (nucleography) thus allowing an accurate interpretation of the form of the nucleus and the changes in a disc

Nutrition of intervertebral disc

The upper and lower surfaces of the body of an adult vertebra are formed of modified spongy bone except at the periphery of each surface where a ring of compact bone represents the fused annular epiphysis (Fig 22a) In an ovoid area which in lumbar vertebrae is situated nearer the back than the front the perforations are smaller and fewer and this more compact plate indicates the site of the nucleus pulposus in the intervertebral disc The porous nature of these surfaces is associated with at least a part of the nutrition of the disc from the vertebral bodies In life, the surface of the bone within the epiphyseal ring is covered by a thin plate of cartilage (Fig 22b) to which the fibres of the annulus fibrosus are attached and it has been debated whether this cartilage plate should or should not be regarded as a part of the disc In this chapter, however the cartilage plates have for descriptive purposes been regarded as parts of the vertebrae

The two components of the intervertebral disc the annulus fibrosus and nucleus pulposus, are avascular except for the most peripheral fibres of the annulus which receive a blood supply from the adjacent vessels, the different reaction of the

importance Puschel (1930), Keyes and Compere (1932) and Schummelfeder and Schummelfeder (1949) all gave essentially similar figures. In the newborn the water content is almost 90 per cent but with the increase in years it decreases so that in the aged it is under 70 per cent. The nucleus pulposus in the young is clearly demarcated from the surrounding annulus fibrosus and on account of its plasticity and the composition of the annulus fibrosus the young vertebral column is endowed with great resilience. An increase in the load in the vertebral column causes a flattening of the nuclear substance and a resultant outward thrust on the annulus fibrosus, and if the increase in load is applied suddenly it allows the disc to act as a hydraulic shock absorber. Furthermore, the semi fluid character of the nucleus permits it to adapt itself readily to changes in its form as one vertebra rocks upon another during movements of the column, and this distortion of the nucleus in turn ensures an even distribution of compression forces over the upper and lower surfaces of the vertebral bodies irrespective of whether the column is in a position of flexion or extension.

Sylvén and his co workers at Stockholm are amongst many who have made notable contributions to the finer analysis of the nucleus pulposus, its nutrition and the changes that occur in it with age. Its avascular nature is generally recognized and there is no real evidence that it is innervated though such a claim has been made. Sylvén (1950) considered that the nucleus may be regarded as a three dimensional honeycomb gel formed by a dense network of interlacing collagen fibrils embedded in an amorphous matrix. These fibrils form the fibres that are shown at relatively low magnification in Fig 21a. In young subjects the fibrils stain like reticulin and are regarded by many as such. They constitute about 50–60 per cent of the total dry weight of the nucleus (Sylvén 1954). The fibrils have a banded structure which is characteristic of all vertebrate collagen with spacings of 640Å. The amorphous matrix surrounding the fibrils according to

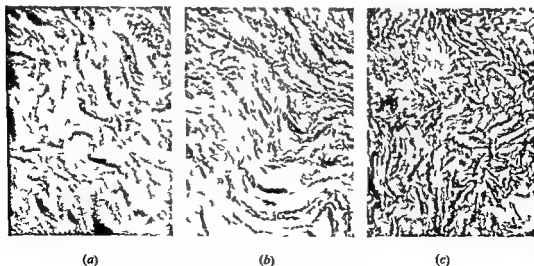


FIG 21—Three sections of the nucleus pulposus at different stages showing maturation of fibrous tissue within the nucleus. (a) Child of 10 years of age showing tapering fibres of elaborated collagen ($\times 55$). (b) Adult of 24 years of age showing denser collagen net work ($\times 55$). (c) Adult of 63 years of age showing dense collagenous mass ($\times 55$). (Reproduced by courtesy of the Editor *Edinb med J*)

references in the literature to disc degeneration in dogs (Hoerlein, 1953, Hansen 1951) Hoerlein in routine autopsies on 130 dogs, found that 41.5 per cent showed disc protrusions and that 63 per cent of all dogs more than 4 years old had lesions

The intervertebral disc between the fifth lumbar vertebra and the sacrum is larger than any other avascular tissue in the body which may partly account for the early degenerative changes that so frequently occur in it

Joints of Luschka

Cave, Griffiths and Whiteley (1955) mentioned the clinical importance of the small synovial joints that lie at the sides of the intervertebral discs of all the cervical vertebrae. The capsule of each of these joints forms the antero-medial wall of the cervical intervertebral foramen and each, it is stated, must restrain the disc from protrusion into the foramen

Longitudinal ligaments

The general features of the anterior and posterior longitudinal ligaments are well known and fully described in standard works

Joints of vertebral arches

The joint between the articular processes or zygapophysial or posterior joint has all the features of a synovial joint with a fairly loose capsular ligament especially in the cervical region which allows appreciable movement between the superior and inferior articular processes of adjacent vertebrae. The set and form of the articular processes is a factor in determining the range of movement permitted between vertebrae but these features of the processes are outside the scope of this chapter. Francis (1955) has made a study of the variations in the articular processes of cervical vertebrae and in analysing the vertebrae of 328 individuals between 25 and 36 years of age concluded that none had a form that would make the zygapophysial joints particularly unstable

The *interspinous*, *supraspinous* and *intertransverse* accessory ligaments of the vertebral joints are all formed of collagenous tissue but the *ligamenta flava* which pass between adjacent laminae are formed of elastic tissue. The elasticity allows the laminae to be separated in flexion of the trunk and on the resumption of an extended position the ligaments retract without the formation of a fold that might press on the dura or be caught between laminae

Intervertebral foramina

The boundaries of the intervertebral foramina are shown in Fig 11. Although the relative size of the structures is different in this foetal specimen from the adult, the decrease in size of the lumbar foramina from above downwards however, is very evident (Dansforth and Wilson 1925) with an associated increase in size of the nerves. Most important of all, the close relationship that exists between nerves and intervertebral discs is obvious. The small upper lumbar nerves are in close relation to the pedicle that bounds the foramen above and are separated from the lower pedicle by numerous venous channels which are present although less numerous in the lower foramina, the posterior boundary of the foramen is formed by the articular processes participating in the posterior joint (zygapophysis)

superficial and deep parts of the annulus to injury (Smith and Walmsley 1951) has been associated with their different mode of nourishment. The presence of vessels in tissues with the physical characters of the annulus fibrosus and nucleus pulposus is incompatible with their function of weight transmission and the avascularity of the disc is characteristic at all stages of its development (Keyes and Compere 1932). The nutrition of the intervertebral disc would appear to be dependent, therefore, on the diffusion of fluid into it from the vertebral bodies and also from the vessels of the peripheral part of the annulus.

Recently, Brodin (1955) has reported his findings in the intervertebral discs of rabbits after the intravenous injection of fluorochrome. The cartilage plates rapidly absorbed the fluorochrome, the contiguous parts of the annulus however, showed a rather weak fluorescence, which decreased gradually as it was traced centrally towards the nucleus pulposus where no fluorescence was observed. This absence of colouring in the nucleus pulposus may be correlated with the statement of Sylven (1950) that nutrition by diffusion represents a very slow method of transportation and that other mechanisms must play a role in the nutrition of the nucleus. Charnley (1952) has postulated that on account of its

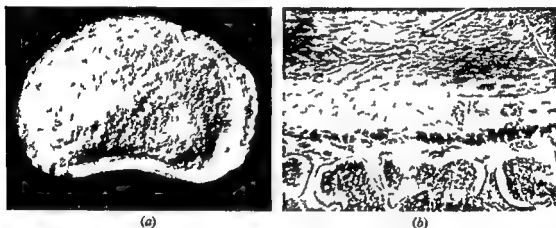


FIG 22 —(a) The upper surface of a lumbar vertebra showing the peripheral fused ring of compact bone (broken off at right side) and within it modified spongy bone. (b) A section through the upper part of a vertebra covering cartilage and nucleus pulposus of a boy of 17 years. The pores in the surface of the bone are apparent and are adjacent to the irregular zone of calcified cartilage which is stained darkly ($\times 16$). (Reproduced by courtesy of the Editor *Edinb med J*)

hydrophilic properties the nucleus may under certain conditions acquire an abnormal amount of fluid and an abnormally high internal pressure and that this may promote an attack of acute lumbago. Sylven also makes reference to the interfibrillar pores within the gel of the nucleus pulposus which will transmit only molecules smaller than 15\AA (1\AA = one tenth of a milli micron).

The degenerative changes that occur in the human discs have frequently been regarded as a normal physiological process associated with their poor nutrition. There is ample evidence that the upright posture is not the sole cause of the degenerative changes that occur in the discs of man for there are numerous

DEVELOPMENT OF NUCLEUS PULPOSUS

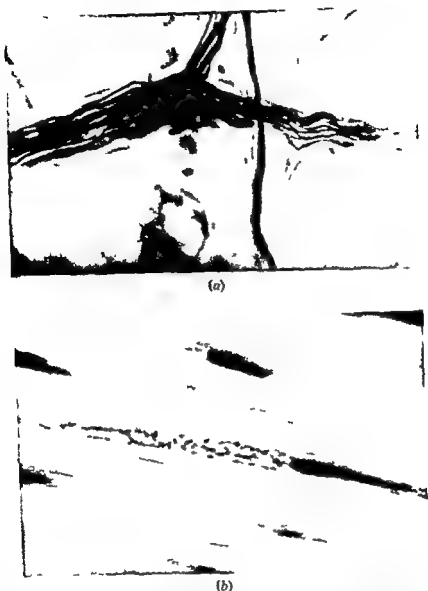


FIG 23 —(a) Nerve fibres of different calibres on the free surface of the anterior (ventral) longitudinal ligament of the dog ($\times 460$)
 (b) Terminal nerve plexus lying within the substance of the posterior (dorsal) longitudinal ligament of the dog. A nucleus of the terminal plexus is seen at its right end but the other dark areas represent fibroblast nuclei ($\times 1125$). Specimens prepared by Dr J H Mulligan and reproduced by his courtesy

sented by either a single or double stems. Pedersen, Blunck and Gardner (1956) state that it contains many myelinated fibres, mostly small, but some exceed 10 microns in diameter, and that probably the smaller ones are pain fibres and the larger ones proprioceptive in function. The nerve passes through the foremost part of the intervertebral foramen close to the lower border of the pedicle and then proceeds towards the mid line giving cranial and caudal branches that supply the periosteum, the posterior longitudinal ligament, the dura mater and the adjacent vessels. The branches of each meningeal nerve overlap the branches of the next

Innervation of the vertebral column

The details of the innervation of the vertebral column assumed practical importance with the appreciation that pathological changes in the intervertebral joints may cause low back pain and sciatica

The formation of spinal nerves within the intervertebral foramina by the union of anterior and posterior roots is a well recognized anatomical fact as is also the paravertebral position of the sympathetic trunk in a more anterior plane. Each spinal nerve is united to the sympathetic trunk by one, two or more rami communicantes. These, along with a multitude of small nerves that arise from them or from the larger nerve trunks which they connect have been described by Stilwell (1956) as the paravertebral plexus. The concept of such a plexus lying between the spinal nerves and the sympathetic trunk may advance the understanding of the innervation of vertebral column structures because the fibres that arise from this plexus may contain both somatic and autonomic components; it is indeed the composite nature of the nerves that has led to difficulty in a functional analysis of the nerve fibres of the vertebral ligaments and joints. The paravertebral plexus however is not a sharply demarcated entity nor is it limited to the sides of the bodies of the vertebrae because fibres from it pass forwards on to the front of the column and also inwards through the intervertebral foramina to the vertebral canal. The site of origin of a nerve fibre is not therefore indicative of its function for the rami contain not only the preganglionic and postganglionic fibres of the autonomic system but also sensory somatic fibres which have their cells in the spinal ganglion. All nerves to the vertebral column to the dura mater and to blood vessels within the vertebral canal irrespective of their origin are therefore regarded as mixed nerves.

The *anterior longitudinal ligament* is richly supplied with fibres which reach it and the adjacent periosteum by passing through the sympathetic chain or, in the cervical region through the vertebral plexus around the artery and vein of that name. The vertebral plexus arises from the stellate or inferior cervical ganglion but Siwe (1931) described the relatively large branches that it receives from the cervical nerves and also from the ninth and tenth cranial nerves. At the lateral border of the ligament most of the nerve fibres enter into its substance but some may course on its free surface (Fig 23a). Stilwell (1956) noted that in the monkey most of the fibres pass towards the deep surface of the ligament and, in the ensuing branching and rebranching extend not only into the next above and next below segment but also pass across the mid line to overlap those of the other side. This extension of vertebral column nerves beyond their own segment appears to be characteristic of vertebral column innervation and is comparable to the overlap that occurs in the zonal innervation of the skin of the trunk by spinal nerves.

The *posterior longitudinal ligament* is supplied by recurrent nerves and at least one of these passes inwards through each intervertebral foramen. This nerve was first described by Luschka (1850) and because some of its fibres pass to the epidural venous sinuses and the bone of the vertebra he named it the *sinu vertebral nerve*. Neither this nor the *NA* term namely meningeal nerve indicates the full distribution of this nerve. Its origin is variable; it usually has one contribution from the spinal nerve or its posterior root and another from one of the nerves of the paravertebral plexus, and in its passage medially it may be repre-

BIBLIOGRAPHY AND REFERENCES

joints and downwards to the lumbo sacral joint From the resting position extension has a greater range than flexion because much seeming flexion occurs at the atlanto occipital and hip joints

To the controversy regarding the axis or axes of movement in flexion and extension of the vertebral column Charnley (1951) has made a notable contribution He recorded that the essential feature of lumbar movement is not so much the location of an axis of movement between each pair of vertebral bodies but rather the 'bending axis' for the spine as a whole, namely the longitudinal axis which undergoes the least changes in length during transition between extreme flexion to extension In this movement, the measurement over the anterior surfaces of the 5 lumbar vertebrae increased by 12 millimetres and that over the posterior surface decreased by 5 millimetres This accords with the radiographic appearance that during flexion of the column the anterior borders of the vertebrae are approximated and the distance between the posterior borders is increased and vice versa during extension The movement Charnley reported is least in a line passing somewhere through the posterior quarter of each intervertebral disc and this he regarded as the bending axis this may be correlated with the eccentric position of the nucleus pulposus in the lumbar discs No doubt, too, as the nucleus pulposus is more centrally placed in the thoracic and cervical regions there is a corresponding forward shift of the bending axes in these regions

The mechanics and dynamics of the vertebral column and the mechanics of posture have been fully considered by Steindler (1955)

It must justifiably appear illogical to undertake any consideration of the column without a simultaneous study of the muscles which not only participate in maintaining the normal alignment of the vertebrae but also produce movement between them The importance of muscles in the mechanism of the column is however fully appreciated and therefore no account is given

The Editors of this volume consider that an up to date, accurate appraisal of the anatomy of the vertebral column is essential Much of what is incorporated in this chapter formed the basis of a Sir John Struthers Lecture which was delivered before the Royal College of Surgeons Edinburgh on 21st May 1952 and subsequently published in the Edinburgh Medical Journal We wish to express our gratitude to the Editors and Publishers of that Journal for their permission to reproduce many of the figures here included The article has indeed formed the basis for this chapter but it has been completely re written and several new illustrations have been added

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above and below by about half a vertebra so that all the innervated structures within the vertebral canal would appear to be supplied by at least two nerves

Dr Mulligan of this Department encountered within the anterior and posterior longitudinal ligaments of dogs many nerve fibres terminating in an extremely fine elongated plexus of fibrils (Fig 23b) often related to several distinct nuclei (1957) Stilwell (1956) stressed the repeated bifurcations of nerves in the longitudinal ligaments and stated that some nerve fibres have 5-15 endings with delicate arborizing branches whose tips taper into invisibility

The joints of the *articular processes* are supplied by twigs from medial branches of the posterior primary rami of the spinal nerve and these also innervate the *interspinous ligaments* but because of the downward inclination of the nerves each interspinous ligament is supplied by the next most cranial segment (Pedersen Blunck and Gardner 1956)

There is ample evidence that stimulation of any of the many nerves to the ligaments or bone of the vertebral column may cause pain, but this may be difficult to locate partly because of the overlap of the nerves and partly because of the multiplicity of innervated structures of the vertebral column

Hirsch (1950) has provoked lumbago like pain by injecting saline solution under pressure into the nucleus pulposus of a degenerated disc. He believes that pressure alone determines the violence of the pain and that the impulses are conveyed by the nerve fibres in the posterior longitudinal ligament, because when this region is anaesthetized, pain does not occur

Movements

The extent and nature of the movements that occur in the different regions of the column are much debated. The mobility between any two vertebrae is certainly dependent on the types of joints between them, the thickness of the intervertebral disc, the set of the articular processes and the articulations with different types of ribs.

At the atlanto occipital synovial joints there is free range of flexion and extension which has been variously assessed between 21.7 degrees (Knese 1948) and 35 degrees (Strasser 1913). The recorded measurements of the range of movement that is permitted at other synovial joints reveal so much individual variation that a single figure is not only meaningless but is also misleading.

The atlanto axial synovial joints which allow a wide range of axial rotation contribute with the atlanto occipital joints one functional unit.

Although the extent of flexion, extension, lateral flexion and rotation vary considerably in the different regions of the column, all the movements can take place in all regions. Thus despite the set of the articular processes, some degree of rotation is possible even in the lumbar region because of the laxity of inter articular joint capsules, but in this region such a movement is always associated with lateral flexion. Capener (1944) has correlated the coronal orientation of the lumbo sacral facets with the probability that a greater degree of rotation may occur here than in the rest of the lumbar region. In the thoracic region rotation can be carried out independently of simultaneous lateral flexion, but on the other hand thoracic lateral flexion is not possible without simultaneous rotation (Farkas 1954). Mobility is noticeably at its least in the mid thoracic region and increases as the column is followed upwards to the axis and atlas.

CHAPTER 2

CONGENITAL ANOMALIES

R I HARRIS

INTRODUCTION

CONGENITAL anomalies of the spine arise from a variety of causes. Some, such as hemivertebrae, fused vertebrae or assimilation of the ribs to the occiput, are due to imperfections in the segmentation of the mesenchyme which under ordinary circumstances would lead to the development of a normal vertebral column. Some, such as Morquio Brailsford chondro osteodystrophy, are due to a generalized congenital disturbance in the development of bone, cartilage and connective tissue. Occasionally a congenital metabolic disorder such as alkaptonuria may cause serious damage to the vertebral column. Intra uterine pathological events may cause imperfect development of the foetus including imperfections in the development of the vertebral column. Finally for convenience there has been included in this section one condition (spondylolisthesis) for which we lack indisputable evidence of a congenital origin though there is suggestive evidence that the disease is familial.

Congenital anomalies of the spine are of clinical importance because of the deformities they cause and because of the consequent interference with function of the spine and thorax. In addition and of particular importance because of its gravity is the disturbance of function of the central nervous system which may accompany the deformity. These neurological problems may be the result of deficiency in the development of the central nervous system itself, which parallels the defective development of the vertebral column (meningocele accompanying spina bifida), or they may be caused by localized pressure upon the pons, medulla, spinal cord or cauda equina produced by the spinal anomaly or by changes secondary to it. Primary deficiencies of development of the central nervous system accompanying spinal anomalies manifest themselves by symptoms and signs present at birth. The resulting disablement may remain unchanged throughout life or it may increase because of additional trauma imposed upon the central nervous system by the deformed spine during growth.

Neurological problems of particular interest are those secondary to damage caused directly by the spinal deformity or to changes in the spine adjacent to the spinal deformity and consequent upon it. The manifestations of this type of neurological damage are seldom present at birth. They develop later in life during growth or when the spinal deformity is increasing or as the result of an injury, or when changes are occurring in the spine secondary to the original congenital deformity. They may appear first in childhood, adolescence or even in adult life. They tend to progress slowly in severity. Examples are stresses upon the cord induced by growth of the vertebral column in the presence of a bony anomaly (diastematomyelia), paraplegia in congenital scoliosis, localized pressure upon the cord by protruded intervertebral discs or osteophytes developing in areas

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CONGENITAL VERTEBRAL ANOMALIES DUE TO IMPERFECT SEGMENTATION

injection of the insulin in terms of foetal development determines the part of the skeleton in which the defect will appear. The action of insulin is most marked in areas of greatest skeletal activity.

Duraiswami's observations may have a clinical application especially in pregnancies of diabetic women receiving large doses of insulin. A suggestive case is illustrated in Fig. 24.



FIG. 24—Radiograph of a 6 month still born foetus whose mother suffered from severe diabetes mellitus necessitating large doses of insulin during pregnancy. Multiple congenital deformities of the skeleton are present including abnormalities of the vertebral column.

CONGENITAL VERTEBRAL ANOMALIES DUE TO IMPERFECT SEGMENTATION

Congenital abnormalities of the vertebral column due to imperfect segmentation (Keith 1933), whether genetic or possibly due to agents acting upon the foetus *in utero*, can best be discussed in relation to the spinal segment in which they occur because it is to this that the symptoms and signs and neurological complications are related.

Fusion (assimilation) of the atlas to the occiput

The atlas is derived from the caudal half of the last occipital sclerotome and part of the first cervical sclerotome. The odontoid process of the axis represents the body of the atlas. This complex origin perhaps accounts for the significant incidence of congenital anomalies of the upper part of the cervical spine. The

adjacent to the congenital deformity because of long continued abnormal stresses upon the first mobile segments above or below an immobile area of the spine choking of the central nervous system as it emerges through the foramen magnum by the upthrust of the odontoid process into its lumen, and at any age injury to a deformed spine may precipitate neurological damage which would not occur in a normal vertebral column

The neurological signs which develop in congenital anomalies of the spine vary in severity. They are often diffuse and complex and difficult to interpret. Not infrequently they lead to the mistaken diagnosis of disseminated sclerosis or syringomyelia. In assessing them consideration must be given to the level of the cord which is subjected to pressure and to the spinal tracts passing through this area. Consideration must also be given to the possible existence of anomalies in the central nervous system itself (Arnold Chiari deformity, diastematomyelia, hidden meningocele). In the last two decades much progress has been made in the elucidation of these neurological problems chiefly by neurologists assisted by the use of improved diagnostic techniques such as myelography, and by neurosurgeons who at operation have the opportunity to demonstrate the pathological lesion and thus make it possible to correlate and interpret history, symptoms, signs and radiographs. The field is one in which the interests and skills of the neurologist, neurosurgeon, radiologist and orthopaedic surgeon overlap. Close co-operation between them is essential for the successful management of these problems. This is especially true when the neurological damage overshadows the severity of the congenital deformity of the spine (paralytic equinovarus deformity associated with spina bifida occulta).

In the past it has been generally accepted that all congenital abnormalities are due to abnormal chromosomes and the frequency with which congenital abnormalities appear in successive generations seems to support this genetic concept of their origin. Isolated examples of congenital deformities have been regarded as mutations in the germ plasma. In the past decade, however, there has appeared increasing evidence that developmental defects can sometimes be the result of intra uterine damage to the foetus by various agents. Since they are present at birth they can correctly be designated congenital defects though they are not due to an inherited chromosome deficiency. The problem of congenital defects due to intra uterine injury was brought conspicuously to our attention by the report of Gregg (1941) upon the occurrence of blindness and deafness in children whose mothers had suffered from rubella in the early weeks of pregnancy. In addition to this evidence that the virus of rubella can cause congenital defects by intra uterine injury there is much experimental work which demonstrates that many agents—x irradiation, hypervitaminosis and hypovitaminosis, hormones, trypan blue (Gillman, Gilbert and Gillman, 1948)—can cause malformation of the foetus. The work of Duraiswami (1952) is of particular interest to orthopaedic surgeons. By the injection of a single dose of insulin into fertilized hens' eggs at an appropriate stage of development he was able to produce a great variety of malformations of the skeleton. Insulin was chosen as the experimental agent because of its effect upon carbohydrate metabolism. The ground substance of bone, being composed in considerable part of mucopolysaccharides, is profoundly disturbed in its metabolism during the period of action of the insulin. During that time a fault in the laying down of the skeleton occurs which is never corrected. The moment of



FIG. 27—Lateral radiograph of assimilation of the atlas to the occiput. The tip of the odontoid process is projected upwards into the foramen magnum above Chamberlain's line (posterior margin of hard palate to posterior margin of foramen magnum).

most common is fusion of the atlas to the occiput—the result of imperfect segmentation of the first cervical sclerotome from the last occipital sclerotome (Figs 25, 26 and 27). The frequency of this anomaly is disputed—the figures quoted by List (1941) range from 0.081 per cent to 2.76 per cent (*see also* McRae 1953).

The fusion of the atlas to the occiput may be complete so that the whole circumference of the atlas is attached to the margin of the foramen magnum or it may be incomplete in any degree. The anterior arch is usually fused, other regions especially the posterior arch, may be incompletely fused or even absent (Figs 25 and 27).

Uncomplicated fusion of the atlas to the occiput probably causes no symptoms and insignificant signs. Few patients suffer disablement sufficient to attract attention.

The demonstration of the lesion is not always easy—good radiographs are essential, and often a sagittal laminogram is of great value (Fig 28).

Three aspects of the deformity must be recognized. Their clinical signs and their treatment are entirely different from one another.

Uncomplicated fusion

The atlas is fused in whole or in part to the occiput and there are no complicating accessory factors. The patient has no great discomfort or disability unless subjected to injury. The changes having been present since birth he may be



FIG 25—Assimilation of the atlas to the occiput . Anatomical specimen . The atlas is fused to the occiput is less thick than normal and is incomplete posteriorly . The facets for the axis are on different planes so that the head must have been tilted to the right



FIG 26—Fusion of atlas to occiput at the atlanto occipital articulations . Anatomical specimen . It is not certain whether this is a congenital abnormality or ankylosis of the articulations secondary to disease but probably the former . Compare with Fig 25 to appreciate the shortening of the neck and tilting of the head which takes place in assimilation of the atlas to the axis . This results in projection of the dens upwards into the foramen magnum (see Fig 27)



Fig 27—Lateral radiograph of assimilation of the atlas to the occiput. The tip of the odontoid process is projected upwards into the foramen magnum above Chamberlain's line (posterior margin of hard palate to posterior margin of foramen magnum)

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FIG 28—Assimilation of atlas to occiput. The laminograms in (a) flexion and (b) extension show the range of luxation of the atlas on the axis. Note also the projection of the odontoid process upwards into the foramen magnum above Chamberlain's line owing to the diminished height of the atlas and the loss of the space of the atlanto occipital articulation. The arrows point to the atlas fused to the occiput.

unaware of anything abnormal. He has no pain or neurological disturbance. All movements of the head on the neck are limited. Because the fused atlas often is asymmetrically developed the head may be tilted or rotated from the midposition (Fig 25). The neck is somewhat short and the hair line low because the joint space between the atlas and occiput is narrow or absent and because the imperfectly developed atlas is thin. The oddly poised head and the hunched shoulders are characteristic. The signs are present from birth, are recognizable early in life and do not increase.

Since the patient is little incapacitated and since nothing can be done to correct the deformity or increase the range of movement, no active treatment is indicated unless injury has precipitated symptoms.

Fusion with forward luxation of the atlas on the axis

Certain cases of fusion of the atlas to the occiput can cause disability because a subluxation occurs between the atlas (fused to the occiput) above and the axis below. All movement occurs at the atlanto axial joint which is subjected to abnormal stresses so that the ligaments, especially the transverse ligament, may be stretched. If suddenly and severely injured they may be torn. Abnormal forward movement of the atlas on the axis can then occur on flexion. The odontoid process is displaced backwards relative to the atlas. In severe cases it may lie in the middle of the ring of the atlas. In this position the tip of the odontoid process can compress the cord against the posterior margin of the foramen magnum, especially if, as is often the case, the odontoid is projected upwards through the foramen magnum.

above the level of Chamberlain's line by the absorption of the atlas into the occiput (Figs 25, 26, 27 and 28)

Trauma often is the factor that initiates symptoms and is the final factor that stretches or ruptures the transverse ligament

The symptoms caused by subluxation of the atlas on the axis are related to the function of the first and second cervical nerves and of the upper segments of the cervical cord and the tracts which pass through it. There is limitation of movement of the head on the neck. Pain at the base of the skull radiating up the back of the occiput. Some degree of quadriplegia is characteristic. If the onset of symptoms is insidious their diffuse and vague character may render diagnosis difficult, and disseminated sclerosis or syringomyelia may be suspected. Clumsiness in walking and numbness in the extremities progressing after injury often are due to this lesion. Good lateral radiographs and laminograms contrasting the changes in position induced by flexion and extension are essential for diagnosis (Fig. 28)

Treatment may be demanded by the severity of the symptoms or their progression. The treatment of choice is posterior fusion of the first and second cervical vertebrae after the deformity has been reduced by extending the head on the neck. Since the atlas is already fused to the occiput this operation in effect is fusion of the spinous process of the axis below to the posterior arch of the atlas and to the occiput above.

Fusion associated with platybasia or with Arnold Chiari deformity or with both

Fusion of the atlas to the occiput may be accompanied by anomalies of adjacent structures and this combination is prone to cause injury to the pons and medulla and disturbance in the circulation of cerebrospinal fluid. The two most frequently complicating anomalies are platybasia and the Arnold Chiari deformity. Neither is a congenital deformity of the spinal column but their frequent association with fusion of the atlas to the occiput and the serious problems which arise from this association necessitate their mention.

Platybasia, or basilar impression, is a deformity of the base of the skull in which the foramen magnum and surrounding area is thrust upwards moulding the base of the cranium into a flat plane or even pushing it farther into the cranium to form an inverted saucer like depression (List 1941). The cause not known with certainty, is probably a developmental weakness of the base of the skull, the weight of the head being sufficient to deform the base of the skull, which is gradually invaginated. The outline of the foramen magnum becomes deformed and its size is diminished so that the medulla may be compressed. At its exit from the cranium the medulla may be sharply angulated around the elevated anterior margin of the foramen. These constrictions cause pressure symptoms related to the pons and the medulla. They also interfere with the free circulation of the cerebrospinal fluid and may give rise to internal hydrocephalus (Figs 29 and 30).

The Arnold Chiari deformity of the brain stem and cerebellum consists of (1) Arnold's deformity, an overgrowth and downward displacement of the inferior part of the cerebellar hemispheres so that the paired tongue like processes hang down through the foramen magnum on the posterior surface of the medulla and upper cervical cord and (2) Chiari's deformity a bulging or kinking of the medulla upon the uppermost segments of the cervical cord as though they were telescoped into each other. Commonly both lesions co exist. Both deformities occupy



FIG 29 —Lateral radiograph (laminogram) of a case of platybasia and Arnold Chiari syndrome. The basilar angle is flattened and the tip of the odontoid process is above Chamberlain's line. In this case there is no assimilation of the atlas to the occiput.



FIG 30 —Myelogram of the case shown in Fig 29. Great defect in the oil column at the cervico occipital level.

space within the lumen of the foramen magnum and may occlude it compressing the pons, medulla and cerebellum and obstructing the flow of cerebrospinal fluid.

When platybasia and Arnold Chiari deformity complicate fusion of the atlas to the occiput, the symptoms of both deformities due to the choking of the brain stem within the foramen magnum are accentuated by upthrust of the odontoid process into the foramen magnum, crowding an already overcrowded space. If in addition the atlas undergoes forward subluxation on the axis, the compression symptoms are greatly increased by the impingement of the tip of the odontoid into the anterior surface of the pons and medulla when the head is flexed. It is this ability of atlanto occipital fusion to accentuate gravely the pressure symptoms of platybasia and the Arnold Chiari deformity that makes their association serious. Symptoms are absent at birth. They appear later, usually in adult life, often after injury. Movement of the head on the neck is limited and the head may be held asymmetrically. The neck appears short. There is pain in the upper part of the neck and the occiput, dizziness, cerebellar ataxia, unsteady gait, weakness of arms and legs and manifestations of increased intracranial pressure. Sensory changes may be mild and diffuse and resemble syringomyelia or disseminated sclerosis.

Treatment of this phase demands relief of pressure by removal of the posterior half of the ring of the atlas and the axis and the posterior margin of the foramen magnum. The therapeutic problem is primarily neurosurgical.

Pro-atlas

The last occipital sclerotome may develop as a partially separate entity (pro-atlas) instead of becoming an integral part of the occipital bone (Gladstone and Erichsen Powell 1951). The result may be a rudimentary vertebra fused to the margin of the foramen magnum resembling somewhat an assimilation of the atlas to the occiput but with a normal atlas and axis below it. Alternatively the only evidence of the pro atlas may be isolated bony nodules interposed between the

FIG 31—Pro atlas laminogram. Above the anterior arch of the atlas is a bony mass attached to the occiput. It represents the anterior arch of the pro atlas or last occipital vertebra. Above the tip of the odontoid is an ossicle which is the rudimentary body of the pro atlas.



(a)



(b)

FIG 32—Lateral radiograph showing pro atlas, absence of odontoid process and luxation of the atlas forward on the axis causing quadriplegia. (a) Extension (b) flexion.

occupy the atlas and the odontoid process. Of these a tiny nodule, sometimes seen in radiographs above the tip of the odontoid process represents the rudimentary body of the last occipital vertebra and a larger nodule above the anterior arch of the atlas the rudimentary anterior arch (Fig 31). This anomaly is rare and of unknown incidence. It is not certain that the presence of a pro atlas alone has any clinical significance. It may modify the freedom of movement between the upper cervical spine and the occiput and thus render the cervico occipital region more susceptible of injury. A pro atlas is often accompanied by other congenital anomalies of the cervico occipital region and this combination can certainly cause symptoms. Fig 32 is the radiograph of the cervico occipital region of a 58 year old man who had slowly developed incomplete quadriplegia. It shows congenital absence of the odontoid process, marked subluxation of the atlas on the axis on flexion and extension, and an ossicle representing the pro atlas above the anterior arch of the atlas.

Separate odontoid process (os odontoideum)

Occasionally the odontoid fails to fuse with the body of the axis (Giacomini 1886) and this may be a source of weakness. Severe trauma can avulse the odontoid process and its displacement may cause neurological symptoms by pressure upon the upper cervical cord. Lesser force may stretch the syndesmosis and give rise to pain.

The discovery by radiography of a separate odontoid process after injury presents the problem of whether the effect is the result of the injury or the result of a congenital failure of fusion. In a fresh fracture the line of separation is irregular and is not corticated. In congenital separation the line is smooth and corticated. In the antero posterior view through the mouth the cleavage is through the base of the process. An oblique fracture line may not be seen. In congenital failure of fusion the plane of separation is easily seen and it is curved upwards in the centre. But the distinction between fracture and congenital separation does not necessarily solve the problem of the relationship of symptoms to injury. Injury may fracture a normal process, it may also avulse a separately developed odontoid process from the body of the axis or stretch the fibrous bond. In either case the symptoms are the result of injury (Figs 33 and 34).

Separate odontoid process may be symptomless. Probably symptoms never develop until the syndesmosis is stretched or avulsed by injury. A single injury promotes a concomitant abrupt onset. The long continued stresses of normal use promote a gradual development. Displacement may be small at first and increase. In the young person the odontoid may fuse in the displaced position.

The symptoms of avulsion or stretching of the syndesmosis are limitation of movement of the head and pain at the base of the skull radiating up into the occipital region. If there is much displacement the head will be carried thrust forward. Lateral laminograms are particularly valuable in delineating the lesion.

Treatment may be necessary for the relief of pain and to stabilize the upper cervical spine. This can be accomplished by posterior fusion of the atlas and the axis with the dens as nearly as possible in normal relationship to the body of the axis.



FIG 33—Separate odontoid process (os odontoideum) Lateral radiograph of the neck of a 10 year old boy who developed symptoms immediately after an accident The well corticated outline of the nodule and its separation from the body of the axis distinguish it from fracture of the odontoid process



FIG 34—Antero posterior view of the case illustrated in Fig 33 It shows the plane of separation between the axis and the base of the odontoid process

Congenital absence of odontoid process

A rare anomaly of the upper cervical spine is complete failure of development of the odontoid process The condition is recognized during investigation of a neck which has suddenly become painful when subjected to considerable stress in athletic activity or in a motor accident and is revealed by an antero posterior radiograph through the mouth The injury often produces subluxation or dislocation of the atlanto axial facets It is conceivable that sudden death could occur from complete dislocation (Fig 35)

Subluxation or unilateral dislocation can usually be reduced by traction Fusion of the atlas to the axis is necessary to prevent recurrence

Klippel-Feil syndrome

Klippel Feil syndrome represents an extensive disturbance of the development of the cervical spine Abnormalities of many or all of the cervical vertebrae occur hemivertebrae fusions of adjacent vertebrae and scoliosis of the cervical spine (Fig 36) The condition described by Klippel and Feil in 1912 was a clinical entity short neck tilted head and stiff neck An extreme disturbance in the segmentation of the cervical vertebrae causes a short deformed cervical spine with



FIG 35 — Congenital absence of odontoid process in a 7 year old girl



FIG 36 — Klippel Feil syndrome hemivertebrae and multiple fusions of of cervical vertebrae

fusion of many vertebrae and imperfect development of others. The head appears set on the shoulders. Webbing of the skin over the lateral margins of the trapezius muscles (pterygium colli) may occur and so may congenital elevation of the scapula (Sprengel's deformity). The deformity is unsightly and movements of the neck and head are grossly limited, but otherwise the functional disturbance usually is slight or negligible. The disability is more aesthetic than functional.

No treatment will alter the primary deformity. The accompanying Sprengel's deformity or pterygium colli can be improved by plastic operations. Late complications usually do not occur but injury may precipitate grave symptoms, sometimes paraplegia because of the great stresses concentrated upon the mobile segments of the cervical spine adjacent to the fused segments.

Hemivertebra

Hemivertebra may occur at any level. A supernumerary vertebral body develops on one side only, being wedge shaped with the apex towards the mid line. One vertebra only may be involved or there may be two or three adjacent hemi-



FIG 37—Hemivertebrae with scoliosis. This 8 year old girl had scoliosis from birth which progressed steadily. Two hemivertebrae are fused adjacent vertebrae are wedge shaped and there are distortions and fusions of the ribs on the concave side of the thorax.

vertebrae partially fused to one another. In the thoracic spine a hemivertebra will carry a rib which makes an extra rib on the involved side. Anomalies of the ribs, particularly fusion of adjacent ribs, often accompany hemivertebrae (Fig 37).

A hemivertebra produces or a series of adjacent hemivertebrae produce a fixed and sharply angulated scoliosis of the involved spinal segment convex to the abnormal side. This is a form of congenital scoliosis.

In the cervical and lumbar segments where movement is greatest the deformity and stiffening produced by a hemivertebra throw abnormal stresses upon the

adjacent mobile segments, with consequent undue wear and tear. These changes are manifested by degeneration of the intervertebral discs and marginal osteophyte formation. These secondary changes give rise to symptoms chiefly backache, which appear first in adult life sometimes after a single severe injury.

In late adolescence scoliosis due to congenital hemivertebra may cause paraplegia. Normal growth of the vertebral column, exceeding the corresponding growth of the central nervous system leads to angulation and flattening of the cord about the spinal deformity (Dewar and McKenzie 1949).

Treatment presents difficult problems. It is true that the deformity and its progression and the disablement often are less severe than might be expected from the degree of malformation. The short extent and the rigidity of the angular deformity caused by a hemivertebra facilitate the development of reverse curves above and below which completely compensate for the congenital angulation. If this is accomplished, the general alignment and balance of the trunk remain satisfactory. This happy outcome however is entirely dependent upon the development of adequate compensatory curves soon after the child assumes the erect posture and is possible only if the scoliosis is short and stable and the spine above and below sufficiently mobile. Fortunately many patients achieve compensation spontaneously. Others may be assisted to accomplish balanced curves by an appropriate brace or corrective plasters.

All too often the magnitude of the primary deformity precludes the possibility of the development of adequate compensatory curves. If the compensatory curves are inadequate growth quickly compounds the deformity by lengthening the arms of the congenital curve thus projecting the weight of the superimposed trunk still farther to one side. Deformity and imbalance progress and may become grossly disabling. Fusions between ribs increase the rigidity of the thorax and add to the difficulty of developing a thoracic curve. Attempts to forestall the gross deformity have been made by direct attack upon the involved segment of the spine in infancy. Parts or all of the interposed hemivertebrae have been excised. The end results were poor but the reports are of some value in demonstrating the feasibility of extensive operations upon the vertebral bodies in infants (Compere 1932, von Lackum and Smith 1933, Wiles 1951). The management of severe deformity is not yet perfected. Some form of operative correction followed by stabilization with bone graft appears to offer the best hope of improvement, but there are grave technical problems still to be solved. The less extensive and less severe deformities can be kept under observation to determine whether or not they are progressive. If necessary they should be treated by spinal fusion after plaster jacket correction.

The type of scoliosis associated with neurofibromatosis will be discussed in the section on Abnormalities due to Generalized Congenital Deficiency in the Formation of Bone Cartilage and Connective Tissue on page 50.

Congenital wedged vertebra

There may be deficiency in the development of the anterior part of a vertebral body so that it is wedge shaped with a forward apex. If several vertebrae are involved there may be also deficiency of the front of the intervening intervertebral discs allowing fusion of the involved vertebrae at their anterior margins (Fig. 38) with consequent kyphosis. The trunk is diminished in height by impairment of



FIG. 38—Lateral radiograph of the spine of an 11 year old boy which shows kyphosis due to anterior wedging and fusion of the bodies of T 10 11 12

growth and by the deformity. These are the only disabilities. Active treatment usually is not indicated.

Congenital deformities of the lumbosacral region

The lumbosacral region is particularly prone to developmental disturbances, which though leading to no great deformity diminish the strength and mobility of that segment of the spinal column most subjected to the stresses of weight bearing and of movement. Here small imperfections often cause disabling symptoms because wear and tear changes promote low back pain and sciatica.

Associated neurological lesions constitute the most serious and disabling manifestation of some lumbosacral anomalies.

Sacralization of the last lumbar vertebra

In imperfect lumbosacral segmentation the last lumbar vertebra is assimilated to the sacrum with varying degrees of completeness. There is a change in structure from that characteristic of a lumbar vertebra to that characteristic of a sacral vertebra. The transverse processes, pedicles and inferior articular facets are transformed into the lateral masses of a sacral vertebra. One or both sides may be sacralized. In the former case one side of the deformed vertebra has the lateral mass of a sacral vertebra and is attached to the sacrum; the opposite side has the transverse process, pedicle, lamina and articular facets of a lumbar vertebra. It articulates with the sacrum but is not fused to it. The sacralized lumbar vertebra may be incompletely fused to the sacrum by a syndesmosis or completely incorporated into the sacrum by bone. When both sides are sacralized and the vertebra

is completely fused to the sacrum by bone the sacrum is lengthened by an extra vertebra at its upper end This form of the anomaly seldom causes symptoms Such as do occur are the result of wear and tear changes in the articulations of the lumbar spine above concentrated upon four lumbar vertebrae instead of being distributed throughout the normal five



FIG 39—Anatomical specimen of unilateral sacralization of the fifth lumbar vertebra Note the osteophytes which have developed on the free side

FIG 40—Radiograph (Hibbs projection) of a patient with in complete unilateral sacralization of the fifth lumbar vertebra (syndesmosis) on the right side He reported because of low back pain and left sciatica



CONGENITAL VERTEBRAL ANOMALIES DUE TO IMPERFECT SEGMENTATION

Sacralization of the last lumbar vertebra gives rise to most trouble when unilateral and incomplete (Figs 39 and 40). The syndesmosis attaching the sacralized half to the sacrum permits some movement, less on the sacralized side than on the normal side. This asymmetrical movement invites wear and tear changes in the apophyseal articulations and the intervertebral disc and these changes cause low back pain and sciatica. Symptoms develop most frequently and most severely on the side which is not sacralized because here there is greater movement which is distorted and therefore greater wear and tear. Similarly there is greater susceptibility to injury.



FIG 41—Trapezoidal body of the fifth lumbar vertebra. This 43 year old woman suffered from backache for many years culminating in left sided sciatica.

Hibbs' antero-posterior radiographic projection is useful in revealing the outline of the fifth lumbar vertebra free of overlapping shadows. The x-ray is directed upwards and backwards through the disc between L5 and S1. It is easiest with the patient recumbent (Fig 40). In certain cases Hibbs' projection with the patient standing though technically more difficult to accomplish gives valuable information with regard to the effect of inequality of leg length on the position of the fifth lumbar vertebra (Fig 42).

Trapezoidal body of the fifth lumbar vertebra

Viewed by Hibbs' technique the body of the fifth lumbar vertebra occasionally is trapezoidal in outline. This may be compensatory to obliquity of the superior surface of the sacrum (or to a short leg on one side if the patient is standing) but

often there is no apparent cause (Fig 41) The effect of the distortion is to throw abnormal strain upon the apophyseal articulations with consequent wear and tear manifested by low back pain and sometimes sciatica

Obliquity of the superior surface of the sacrum

For obscure reasons the sacrum may develop with its surface higher on one side than on the other This is most clearly demonstrated by an antero posterior radiograph directed obliquely upwards through the disc between the body of the fifth lumbar vertebra and the top of the sacrum If an additional projection of this type is taken with the patient standing any tilt of the pelvis due to difference in length of the two legs can be determined and true obliquity of the superior surface of the sacrum assessed (Fig 42)



FIG 42—Obliquity of the superior surface of the sacrum The patient is standing the legs are of equal length and the x ray is directed upwards through the intervertebral disc between L5 and S1 (Hibbs projection) Because the top of the sacrum is higher on the right side than the left the lumbar spine is directed to the left This is compensated by a lateral curve convex to the left

Obliquity of the superior surface of the sacrum provides a sloping foundation for the vertebral column The compensatory lateral curve produces asymmetrical stresses which in time wear out the apophyseal articulations and the lumbosacral intervertebral disc The body of the fifth lumbar vertebra is moulded into a trapezoidal shape its margins are prolonged into osteophytes the disc space between the body of the fifth lumbar vertebra and the sacrum is thinned and the patient suffers from low back pain and sciatica

If it were possible to detect this asymmetry in infancy the consequent troubles might conceivably be prevented by an appropriate lift to the shoe on the low side Most cases are not discovered until the patient is suffering from low back pain By that time relief can be obtained most effectively by measures which diminish lumbosacral movement A brace may give great comfort but in severe cases and especially those with sciatica from root pressure of a bulging disc operation is expedient (exploration of the lumbosacral disc and lumbosacral fusion) It alone gives hope of complete and permanent relief

Anomalies of structure in the lumbosacral apophyseal facets

Occasionally an inferior facet of the fifth lumbar vertebra is completely absent (Fig 43). The instability from this one sided defect places asymmetrical strains on the lumbosacral joint which lead to abnormal wear manifested by attacks of low back pain.



FIG 43 —Absence of right inferior facet of fifth lumbar vertebra spina bifida occulta of first sacral vertebra. This 21 year old nurse had suffered from recurring attacks of acute low back pain since the age of 10 years. (Radiograph viewed from behind forward)

Absence of both inferior facets of the fifth lumbar vertebra has not been recorded. Something similar is known and causes a special kind of spondylolisthesis. In these cases the inferior facets of the fifth lumbar vertebra and the superior facets of the first sacral vertebra are so imperfectly developed that at some stage the former overslide the latter and dislocation occurs. Once the joint is unlocked there is nothing except the resistance of the intervertebral disc to prevent forward displacement of the fifth lumbar vertebra on the sacrum. The disc slowly gives way and spondylolisthesis develops. It is distinguished from the more common type of spondylolisthesis by the integrity of the neural arch which can be determined by radiograph and at operation.

More subtle than structural changes in the lumbosacral apophyseal articulations are those variations in relationship of structurally normal facets which can impair the strength and stability of the lumbosacral joint. They are important because of the symptoms which arise from them. If the plane of the lumbosacral apophyseal articulation is sagittal it cannot as effectively resist forward displacement of the fifth lumbar vertebra on the sacrum as when the plane of articulation is oblique or coronal. Such joints are unstable and give rise to increased stress. If one joint is disposed in the sagittal plane and the opposite in the coronal plane asymmetrical stresses arise causing wear and tear changes and consequent low back pain.

Spina bifida

Spina bifida (Fig 43) probably never causes symptoms, except through the frequently associated abnormalities of the central nervous system. The commonest of these is meningocele. This may be small and completely enclosed within the spinal canal or it may bulge through the spina bifida and reach a great size in which case some part of the central nervous system protrudes with the meninges to form a meningocele. Spina bifida may occur and cause neurological disturbances at any level in the spinal column but the lumbosacral region is much the commonest.

The neurological symptoms often associated with lumbosacral spina bifida are impairment of sensation in the feet, paralysis or weakness of muscles especially those motivating the foot and toes, imperfect control of bladder and rectum, deformities of the lower extremity, especially of the feet from muscle imbalance. Sometimes the symptoms increase during growth. These symptoms if present from birth and without a clear history of any neurological disease should suggest spina bifida and prompt a search for evidence such as a swelling of the lumbosacral region (meningocele), pigmented hairy skin and radiographic demonstration of spina bifida. Finally a myelogram should be taken. Early detection of the lesion, especially if there is progressive neurological deterioration from an expanding meningocele, will sometimes permit operative relief of pressure and improvement in symptoms.

Hidden meningocele

Hidden meningocele deserves a brief mention because it is associated with a defect in the sacrum which may be congenital. An oval excavation in the anterior wall of the sacral canal is occupied by a meningocele. The defect in the sacrum can be revealed by radiography but myelography is necessary to display the meningocele. There is no associated spina bifida (Fig 44).

Diastematomyelia (diplomyelia)

Diastematomyelia is an anomaly of the spinal column with an associated defect of the spinal cord and meninges. Though rare this lesion is of serious importance because of the accompanying disturbance in function of the spinal cord (Herren and Edwards 1940; Neuhauser, Wittenborg and Dehlinger 1950).

The basis of the lesion is a spur, usually bony, which projects backwards from the body of a vertebra. The spur may also be cartilaginous or fibrocartilaginous. Its apex is attached behind to the dura or to a neural arch. This spur is a barrier to the normal development of the spinal cord which consequently is laid down on either side of it. In fully developed diastematomyelia the spinal cord and its meninges are perforated by the spur and surround it. The spinal canal is widened to accommodate the increased bulk of the duplicated cord and its membranes, as witness the widened interpeduncular distance. The separation of the pedicles from one another is produced by spreading apart rather than by erosion as is the case in spinal cord tumour. The common site is the lumbar or lower dorsal spine (Figs 45 and 46).

If the spur is bony it can be seen in the radiograph but often it is chiefly cartilaginous or fibrocartilaginous. Myelography yields most information demonstrating the bifurcated dural tube surrounding the bony spur.



(a)



(b)

FIG 44 —Hidden meningocele
(a) Defect in the anterior wall
of the sacral canal (b) myelo
gram demonstrates that the
bony defect is occupied by a
meningocele. The patient has
also spondylolisthesis of the
fifth lumbar vertebra

The defect in the spinal cord is manifested by motor and sensory disturbances of varying severity. Somewhat as in spina bifida occulta a child, early in life, is discovered to have a mild foot deformity and some minor difficulty in walking. Later sensory disturbances may be found and bladder dysfunction may become



FIG 45 — Diastematomyelia. The shadow of the bony spur is superimposed on the body of the third lumbar vertebra. The interpeduncular distance is widened from L1 to sacrum and is greatest at L3 and L4.



FIG 46 — Myelogram in a case of diastematomyelia. The radio-opaque oil flows around the spur in the two limbs of the divided dural tube.

apparent. Characteristically the symptoms increase during rapid growth, from the drag upon the spinal cord by the transfixing bony spur. This drag may be so great as to choke the foramen magnum with the pons and medulla and cause internal hydrocephalus.

Improvement can be obtained by removal of the spur. Diagnosis, treatment and rehabilitation call for close association of the neurologist, radiologist, neurosurgeon and orthopaedic surgeon.

ABNORMALITIES DUE TO GENERALIZED CONGENITAL DEFICIENCY IN THE FORMATION OF BONE, CARTILAGE AND CONNECTIVE TISSUE

Any generalized congenital deficiency in the formation of bone and cartilage can involve the spine and lead to its deficient development. Recognition is usually



(a)



(b)

FIG 47—The spine of a 31 year old man with Morquio Brailsford chondro-osteodystrophy (a) Dorsal spine (b) lumbar spine

not difficult because of obvious abnormalities commonly found in the extremities. Four examples may be cited.

Morquio-Brailsford chondro-osteodystrophy

Morquio Brailsford chondro-osteodystrophy is a disturbance of bone formation chiefly in the epiphyses. It is often familial. The early manifestations are difficulty in walking, a stooped posture and kyphosis. The epiphyses of the long bones are developed from irregular centres of ossification and are misshapen. The development of the vertebral bodies is abnormal, especially epiphyseal growth. The vertebral bodies are narrowed anteriorly, not as a wedge but like a cushion with converging surfaces. The spine is shortened and kyphosis is common. As bone deposition progresses to maturity the vertebral bodies form shallow structures, broad from side to side, long from front to back, and with irregular surfaces above and below (Fig 47). The disability from the spinal changes is less than that from the changes in the epiphyses of the long bones, particularly those of the lower extremity.

Fragilitas ossium

Fragilitas ossium produces spinal deformity because of the frailty of bone structure. Superimposed weight compresses vertebral bodies and distorts the alignment of the spine. The height of the patient is greatly diminished by compression. Changes are even more marked in the extremities, particularly the legs, where multiple fractures and distortions diminish their length and disturb their alignment (Fig 48).



FIG 48—The spine in a 39 year old woman with severe fragilitas ossium from birth

Oxycephaly (acrocephaly)

Oxycephaly (acrocephaly) is characterized by fusions of many bones normally separate. The most conspicuous and disabling feature arises from fusion of the coronal, sagittal and lambdoid sutures of the skull, so that the cranium becomes short from back to front, the vault high and pointed, hence the name oxycephaly (pointed skull) or acrocephaly. Fusion of other bones also occurs, notably in the tarsus and metatarsus, carpus and metacarpus. Syndactylism is also a feature.

Fusions of vertebrae may occur—often of the spinous processes and laminae as well as of the bodies. Distortion of the bodies, hemivertebrae or partial fusions are not uncommon. Shortening of the spine and the legs causes dwarfism. Spinal movements are limited.

The structural and functional changes in the spine are not the most important of the disabilities of this disease. The distorted and inexpandible cranium with its secondary exophthalmos and mental impairment and the short extremities with extensive syndactylism are far more disabling.

Neurofibromatosis

Neurofibromatosis (von Recklinghausen's disease) frequently causes a distinctive type of scoliosis. McCarroll's (1950) comprehensive review of 46 cases of neurofibromatosis demonstrated that next to café au lait spots (33 cases) scoliosis (19 cases) was the most frequent clinical and skeletal manifestation of the disease.

The scoliosis may be mild or severe. It is the result of wedging of two or three adjacent vertebrae from deficiency in growth on one side of the involved vertebral bodies. The consequence is a short, sharply angulated and rigid curve which tends to progress because of the persistence of unilateral growth deficiency.

CHANGES DUE TO CONGENITAL DISTURBANCES OF METABOLISM

Adequate compensatory curves above and below are difficult to develop so that deformity is often severe and resistant to treatment (Fig 49)

Other congenital disturbances of bone development may each exert its distinctive influence upon the vertebral column

FIG 49 —Neurofibromatosis and scoliosis. Wedged vertebrae due to deficiency of growth cause a stiff and progressive scoliosis difficult to correct or to compensate with adequate secondary curves



CHANGES DUE TO CONGENITAL DISTURBANCES OF METABOLISM

Alkaptonuria

Alkaptonuria is an inherited defect in the metabolism of certain amino acids especially tyrosine and phenylalanine. Homogentisic acid the end product of this imperfect metabolism is excreted in the urine. The urine turns dark on standing owing to oxidation of the homogentisic acid. Homogentisic acid has an affinity for connective tissue and especially for cartilage with which it combines everywhere in the body. Subsequent oxidation of the homogentisic acid stains black the cartilage of ears, nose, joints and intervertebral discs. The union of homogentisic acid with cartilage causes the ground substance to deteriorate. The cartilage becomes brittle and easily fragmented. In weight bearing joints such as the hip and knee this leads to gross degenerative changes.

In the spine the intervertebral discs accumulate homogentisic acid and become dark by its oxidation. The normal quality of the discs is lost. They diminish in height and lose their elastic qualities. The vertebral column shortens by loss of the disc spacing. The lumbar spine flattens. Later calcium is deposited in the intervertebral discs in an irregular fashion. The spine becomes rigid presenting a clinical picture somewhat resembling that of ankylosing spondylitis. The radiological picture (Fig 50) however, is very different. The intervertebral disc spaces are diminished, the lumbar lordosis is flattened and there is moderate anterior



FIG 50—Lateral radiograph of lumbar spine of a 40 year old man with alkaptonuria. The intervertebral discs are thin and calcification has occurred. The vertebral bodies show marginal lipping and are displaced backwards on the subjacent vertebra.



FIG 51—Skeletal changes in hypophosphatasia. These are most marked in the skull and metaphyses but the vertebral column is also involved. The illustration shows the vertebral column in a child aged 1½ years.

lipping of the margins of the vertebral bodies. There is no involvement of the sacroiliac joints or bony bridging across the intervertebral discs such as occurs in ankylosing spondylitis.

The disease is uninfluenced by treatment. The spinal changes though marked, are less disabling than are those of the diarthrodial joints, especially the hips and knees. There is relatively little pain referable to the spine owing to its rigidity.

Hypophosphatasia

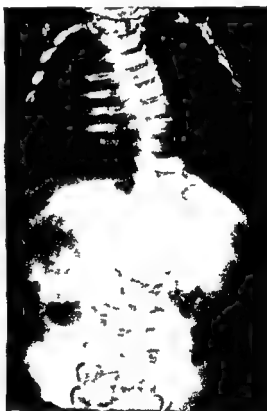
Recent observations on hypophosphatasia by Rathburn (1948), Sobel and his colleagues (1953), Engfeldt and Zetterstrom (1954), Fraser, Yendt and Christie (1955), Fraser and Laidlaw (1956) demonstrate that children can inherit a metabolic anomaly characterized by diminished alkaline phosphatase activity in the blood, a generalized deficiency in skeletal development and the presence of an abnormal amino acid in the urine and blood plasma. There is evidence that the defective development of the skeleton is the result of disturbance in the metabolism of bone matrix. The skeletal changes are most marked in the skull and extremities but there is involvement of the spine also (Fig 51).

The bony changes bear some resemblance to rickets with an element of frailty in bone structure reminiscent of fragilitas ossium. It would be interesting if fragilitas ossium were ultimately proved to be due to an inherited metabolic defect.

INFANTILE IDIOPATHIC SCOLIOSIS

There has been increasing recognition especially in Great Britain of a characteristic type of scoliosis which is present in the first year or two of life. It is recognized fairly often in the first few months and can be recognized at birth. Probably more cases would be recognized at birth or in the first few weeks if routine examination of the new born child was conducted with the possibility of this deformity in mind. The fact that at least some cases of this deformity are present at birth, coupled with Denis Browne's hypothesis (1956) that the deformity is caused by persistent malposture *in utero*, justifies its inclusion in this chapter. It is interesting to recall that Whitman (1917) stated that congenital lateral curvature may occur in infants otherwise normal due apparently to a constrained attitude before birth.

FIG 52—Infantile idiopathic scoliosis in a 4 month old girl. This curve was recognized shortly after birth.



The deformity is said to occur chiefly in boys; the curve is centred at the sixth to the ninth thoracic vertebrae and usually is convex to the left (Fig 52). At birth the lateral curvature of the spine may be so slight that it cannot be recognized with certainty by clinical examination. What can be recognized at birth are transverse wrinkles in the skin about the trunk posteriorly at the right costal margin and the difficulty the child has in bending to the left compared with the ease with which the trunk is bent to the right.

Denis Browne (1956) postulated that the deformity is initiated *in utero* by persistent maintenance of the trunk in a position of lateral curvature. He compared it to his concept of intra uterine malposition of the foot in the development of congenital equinovarus and believed that persistent malposition not only moulds the spine into a lateral curve but also by compression of the lateral margins of

the vertebral bodies on the concavity of the curve damages their capacity for growth on that side and causes the deformity to progress after birth or to recur after improvement. While this hypothesis does not explain the nearly exclusive incidence in males in the mid thoracic region and the preponderance of the convexity to the left it is a provocative idea which deserves further inquiry.

Certain examples of the deformity disappear spontaneously usually before the child reaches 2 years of age but sometimes regression continues beyond that age (Scott and Morgan 1955 1956 and Scott, 1956). Usually, however the case of infantile idiopathic scoliosis which persists beyond 2 years of age or is not recognized until after that age has a grave tendency to progress to a scoliosis of great severity with much rotation and kyphosis. There is severe interference with respiratory and cardiac function because of the deformity and early death is frequent. Braces and plaster have little influence and even early fusion is far from satisfactory. It is a most grave type of scoliosis. If treatment is to be more successful, early recognition and early treatment will be important.



Fig 53—Spondylolisthesis of fifth lumbar vertebra. The characteristic defect is interruption of the neural arch on either side at the pars interarticularis. The anterior fragment consists of body, pedicles, transverse processes and superior facets; the posterior fragment of laminae, spinous process and inferior facets.

SPONDYLOLISTHESIS

Though spondylolisthesis cannot with certainty be regarded as a congenital lesion of the spine it can most conveniently be included in this chapter.

Spondylolisthesis develops in consequence of a structural defect in a vertebra usually the fifth lumbar. The defective vertebra is separated into two parts by a bilateral defect in the pars interarticularis of the neural arch. The fragment of the vertebra is composed of the body, pedicles, transverse and superior articular facets; the posterior fragment includes the spinous process, the laminae and the inferior articular facets. The two fragments are joined at the pars interarticularis by fibrous tissue instead of being solid bone (Fig 53).

We have no certain knowledge of the manner in which the defect develops. It is not congenital. None of the series of examinations which have been carried out upon the spines of newborn children has yet yielded a single example of spondylolisthesis (Batts 1939). It is not the result of a defect in normal ossification. The primary centres of ossification for a vertebra are three: one for the body and one for each half of the neural arch. The latter develop each into one half of the neural arch from the point where the base of the pedicle is attached to the vertebral body to the tip of the spinous process. Each lateral centre, therefore, forms pedicle, superior articular facets, pars interarticularis, inferior articular facets, lamina, and half of the spinous process. At birth the three centres of ossification are still separate. They fuse during the first year. Though the structural defect of spondylolisthesis has not been observed at birth, it sometimes does occur early in life. Kleinberg's (1934) patient, aged 17 months, is the youngest patient the present writer has been able to find.

The fifth lumbar vertebra is most often affected, the remaining lumbar vertebrae with diminishing frequency as they ascend. In a personal series of 217 cases the distribution was L 6 - 1 case, L 5 - 196 cases, L 4 - 16 cases, L 3 - 4 cases. Occasionally more than one vertebra is involved: once in the writer's series of 217 cases two vertebrae (L 4 and L 5) each had the defect. Occasionally the lesion is seen elsewhere than in the lumbar spine: usually in the cervical spine and usually after trauma. The defect may be unilateral in which case the deformity and the symptoms are both less severe than when the defect is bilateral (Figs 54 and 55).

There is some evidence that the disease is familial. Baker and McHolick (1956) examined by radiograph the lumbosacral region of 400 schoolchildren in the first grade (6-7 years of age). Eighteen (4.5 per cent) showed the defect in the neural arch. They were able to examine 29 parents of these 18 spondylolisthetic children. Amongst them the incidence of the defect was 27.6 per cent, which is more than five times greater than the incidence in the general population. Friberg (1939) reported on three generations of the descendants of one spondylolisthetic man. There were 15 cases of spondylolisthesis amongst 66 individuals. Its occurrence in siblings is sufficiently frequent to be significant. Since the defect is absent at birth the inherited factor may conceivably be a weak pars interarticularis which at some early period in life breaks apart under the stress of normal use. This would be a type of stress fracture. In most instances such a fracture would unite. Those that go on to non-union would constitute the cases of spondylolysis and spondylolisthesis which are recognized clinically and radiologically.

The incidence varies throughout the world. In North America the defect is found in 5 per cent of the skeletons in anatomical departments (Willis 1931, Harris 1951). There is some evidence to indicate that it is higher in certain races or occupations: up to 40 per cent in Alaskan Eskimos (Stewart 1931, 1932, 1935).

The fibrous bond that bridges the defect in the neural arch carries much of the stress of weight bearing and movement. In some cases it does this efficiently for many years or even for the lifetime of the patient. Such patients have no symptoms and no deformity and their defect is only discovered by accident. The known 5 per cent incidence of spondylolisthesis and spondylolysis in the material in anatomical museums so greatly exceeds the number of patients who appear for treatment because of their symptoms that many cases must be symptomless and therefore

the vertebral bodies on the concavity of the curve damages their capacity for growth on that side and causes the deformity to progress after birth or to recur after improvement. While this hypothesis does not explain the nearly exclusive incidence in males, in the mid thoracic region and the preponderance of the convexity to the left, it is a provocative idea which deserves further inquiry.

Certain examples of the deformity disappear spontaneously usually before the child reaches 2 years of age but sometimes regression continues beyond that age (Scott and Morgan 1955 1956, and Scott 1956). Usually however the case of infantile idiopathic scoliosis which persists beyond 2 years of age or is not recognized until after that age has a grave tendency to progress to a scoliosis of great severity with much rotation and kyphosis. There is severe interference with respiratory and cardiac function because of the deformity and early death is frequent. Braces and plaster have little influence and even early fusion is far from satisfactory. It is a most grave type of scoliosis. If treatment is to be more successful early recognition and early treatment will be important.

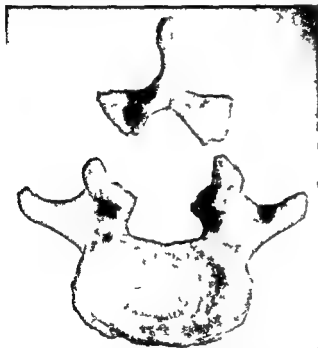


FIG 53—Spondylolisthesis of fifth lumbar vertebra. The characteristic defect is interruption of the neural arch on either side at the pars interarticularis. The anterior fragment consists of body pedicles transverse processes and superior facets; the posterior fragment of laminae spinous process and inferior facets.

SPONDYLOLISTHESIS

Though spondylolisthesis cannot with certainty be regarded as a congenital lesion of the spine it can most conveniently be included in this chapter.

Spondylolisthesis develops in consequence of a structural defect in a vertebra usually the fifth lumbar. The defective vertebra is separated into two parts by a bilateral defect in the pars interarticularis of the neural arch. The anterior fragment of the vertebra is composed of the body pedicles transverse processes and superior articular facets; the posterior fragment includes the spinous process the laminae and the inferior articular facets. The two fragments are bound together at the pars interarticularis by fibrous tissue instead of being solidly fused by bone (Fig 53).

and downward across the sloping upper surface of the first sacral vertebra, thereby initiating the deformity of spondylolisthesis.

The displacement of the involved vertebra may be slight or great. The body of the fifth lumbar vertebra may be completely displaced from the top of the sacrum and lie with its inferior surface applied to the inferior surface of first or second sacral vertebrae. Such gross displacement, however, is rare. More commonly the body of the fifth lumbar vertebra remains on the top of the sacrum but displaced forward a varying amount. In the great majority of cases the displacement is less than half the width of the top of the sacrum. In a personal series of 160 cases of spondylolisthesis of the fifth lumbar vertebra in which the degree of displacement is recorded the distribution was as follows: (1) defect in neural arch present but without displacement of the vertebral body (spondylolysis), 60 cases 37.5 per cent, (2) slight to moderate displacement of the body of the fifth lumbar vertebra (not more than one third the width of the top of the sacrum) 83 cases 52 per cent, (3) displacement greater than one third the width of the top of the sacrum 13 cases 8 per cent, (4) vertebral body completely off the top of the sacrum 4 cases 2.5 per cent.

The factors that determine the amount of displacement of the involved vertebra, the rapidity with which it develops and the uniformity of progression are obscure. Probably the deformity often does not progress at a uniform rate. In the early stages there may be uniform and relatively rapid progression which later slows or ceases by the development of elements of stability. One such is the production of a wedge of bone at the antero superior corner of the body of the first sacral vertebra, and this becomes a buttress to support the displaced body of the fifth lumbar vertebra (Fig. 56). There is not often an opportunity to follow the development of a spondylolisthetic deformity over a period of years, but probably the involved vertebra, even though defective, still has considerable strength and stability. These factors vary from case to case, in individual cases they tend to deteriorate with age. Even so, many cases are strong and stable enough to get through life without symptoms or deformity. Other cases give way but the degree of weakness varies, sometimes little, sometimes great, so that the resulting symptoms and deformity vary greatly.

There have been no recent advances in our knowledge of the aetiology of spondylolisthesis. We are in need of extensive studies of the spine at birth and in the first few years of life. Post mortem specimens for study are also much needed and they are not easy to obtain. Advances have, however, been made in diagnosis and treatment.

Spondylolisthesis manifests itself by certain signs and symptoms and a characteristic deformity. The deformity is the result of the forward displacement of the body of the involved vertebra. It carries with it all the vertebrae above it, leaving behind its own spinous process. Consequently this projects prominently in relation to the spinous processes above it, forming a step kyphosis in contrast to the angular kyphosis of Pott's disease. The pelvis is rotated about a transverse axis passing through the hip joints so that the anterior superior spines of the ilium are raised to the same level as the posterior superior spines or even higher, a remarkably constant sign in spondylolisthesis even when the vertebral displacement is slight. It occurs also in certain lumbosacral disc lesions, but in combination with the step kyphosis and other signs of spondylolisthesis it is a valuable aid to diagnosis.

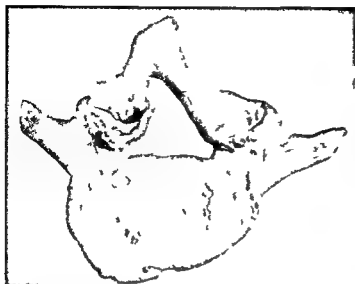


FIG 54—Anatomical specimen showing unilateral spondylolisthesis defect at the pars interarticularis on the right side only. Note the distortion in the outline of the neural arch and the resulting diminution in size of the spinal canal.



FIG 55—Laminograms—right and left oblique projections—of a case of unilateral spondylolysis. The defect is in the pars interarticularis on the left side.

unrecognized. In other cases the syndesmosis gives way under daily stress. The clinical syndrome of pain and deformity then develops. Injury fairly often is the final force which disrupts and stretches the syndesmosis.

When the involved vertebra is the fifth lumbar, as so often happens, the disruption of the syndesmosis throws all the stress of weight bearing and movement upon the intervertebral disc and the longitudinal ligaments. Lacking the normal support of an intact neural arch and the imbrication of the apophyseal facets, these in turn give way and permit the body of the fifth lumbar vertebra to slide forward.

related to the distribution of the fifth lumbar root. This type of root pressure often is bilateral. The distortion of the intervertebral foramen may become so great as to interrupt completely the impulses transmitted through the fifth lumbar root. This causes marked sensory changes and great weakness of the peronei and the dorsal flexors of the toes. The second mechanism producing root pressure involves the first sacral root, usually on one side. It is the result of protrusion of the disc or often of pressure by the irregular osteophytic margins about the defect in the neural arch. Certainly pre-operative myelograms do not consistently show disc protrusions in spondylolisthesis. This form of sciatica based upon first sacral root pressure manifests itself by sensory changes in the outer side of the calf and heel, absence of the ankle jerk and sometimes weakness of the plantar flexors.

It should be emphasized that movement is an important agent in the production of the pain in spondylolisthesis. Even the distortions of structure just described which lead to sciatica first cause pain only when movement makes the narrowed foramen bulging disc or bony spicule impinge upon a nerve root. The structural changes provide the background for sciatic pain; movement is the last factor necessary to bring the structural changes into operation as pain producing mechanisms. Usually this remains true throughout the whole course of the disease. Consequently rest from movement and freedom from weight bearing form the principles of treatment. Only occasionally is the compression of the nerve root by disc or osteophyte or narrow foramen so severe that nerve function cannot be restored by rest.

Spondylolisthesis probably exists often without symptoms. It would be difficult otherwise to explain the discrepancy between the 5 per cent incidence of the defect in adult skeletons in anatomical museums and the much smaller number of the general population who appear for treatment because of symptoms. Certainly it is true that many patients appear first for treatment in the middle or later decades of life yet they probably have had the defect since early childhood.

There is a form of spondylolisthesis that is caused by a different defect of the involved vertebra. In this form the inferior facets of the fifth lumbar vertebra are small or badly disposed in relation to the superior facets of the first sacral vertebra, so that under stress the fifth lumbar vertebra may become unlocked from the first sacral vertebra and slide forward. This really is a dislocation. The clinical symptoms and signs are very like those of spondylolisthesis due to a defect in the neural arch but the essential pathology is different. Two per cent of a personal series of over 200 cases belong to this group.

The diagnosis of spondylolisthesis is assisted by good radiographs. Laminography can help greatly, in the lateral projection, and particularly in the oblique projections which provide the only technique that will permit the certain demonstration of a unilateral defect of the neural arch (Fig. 55). Myelography is of less positive assistance but probably it should be used routinely, as the only means of demonstrating an associated neurological defect (*see under hidden meningocele* on pages 48-49).

The treatment of spondylolisthesis demands sound judgment. Perfect fixation of the involved segment by itself will give complete relief. But this cannot be achieved except by lumbosacral fusion which is no mean procedure. Consideration must be given to the age of the patient, the sex (females can adapt themselves more easily to the restriction of conservative treatment of spondylolisthesis) the

The rotation of the pelvis about its transverse axis may be so great that the thighs are not in a straight line with the trunk even when the hips are fully extended. Hence the patient must stand with his trunk thrust forward if the legs are vertical, or with the hips and knees flexed if the trunk is held erect. The trunk is shortened by the downward displacement that accompanies the forward slipping. Consequently the ribs approach or overlap the iliac crests and transverse creases appear about the waist.

Pain is the symptom that brings to attention most cases of spondylolisthesis. It occurs as low back pain, sometimes with sciatica from root pressure. This may involve either one side or both. Many patients with spondylolisthesis are however free from sciatica.



FIG. 56—Lateral radiograph of spondylolisthesis of the fifth lumbar vertebra with severe displacement. It shows the buttress which develops at the antero superior corner of the sacrum which in certain cases provides some support to the displaced vertebral body.

Low back pain is caused by the instability of the involved vertebra and its forward displacement, both of which put abnormal stresses upon the intervertebral disc and the spinal ligaments. The low back pain is accentuated by movement of the back and by the erect position, either sitting or standing. Recumbency gives quick relief.

Sciatica accompanying spondylolisthesis of the fifth lumbar vertebra is due to pressure either upon the fifth lumbar root or the first sacral root. At least two mechanisms cause this root pressure. The first occurs when the displacement of the vertebral body is considerable. Then the intervertebral foramina between the fifth lumbar and the first sacral vertebrae are distorted and narrowed. In time the narrowing of the foramina compresses the emerging fifth lumbar roots and the compression is accentuated by movement between the fifth lumbar vertebra and the sacrum. In consequence sensory and motor disturbances develop.

of the roots through the intervertebral foramen. Relief of the most severe symptoms may be justification for its use, especially in older patients. The residual symptoms may not be severe and can be accepted with some modification of the patient's way of life. It is true also that a certain degree of stability develops in many cases of spondylolisthesis by the growth of a buttress at the antero superior corner of the body of the first sacral vertebra which supports the displaced body of the fifth lumbar vertebra. Increased experience may allow selection of the cases that will obtain most benefit from this operation. One possible objection to the operation is the difficulty in securing fusion of the lumbosacral region should this subsequently be considered necessary. However, the experience of Bosworth and his colleagues (1955) perhaps indicates that the difficulty in fusing the lumbosacral region after removal of the neural arch may be more theoretical than practical for they have adopted, as their standard procedure for the operative treatment of spondylolisthesis, removal of the loose posterior segment of the neural arch plus lumbosacral fusion with an iliac H graft.

It is one thing to accept the principle that successful fusion of the lumbosacral region is the best way to treat spondylolisthesis and another to describe a simple and certain method by which this can be accomplished. Lumbosacral fusion is technically the most difficult of all spinal fusions to accomplish successfully. It cannot be approached lightly. Nothing but the most exact technique will achieve success in the highest percentage of cases.

The following principles can be laid down as the outcome of experience with over 200 cases (Harris 1951). (1) The fusion should be a posterior fusion uniting the spinous processes and laminae of the involved vertebra to normal vertebrae above and below. Anterior (transperitoneal) fusion of the bodies is impracticable, there are risks of intraperitoneal complications and it is technically difficult to approach the interspace between the fifth lumbar and the first sacral vertebrae if there is more than slight displacement. Posterior interbody fusion (Cloward 1953) has not been used in spondylolisthesis enough to be certain of its merits, if any. The plane of the intervertebral disc between the fifth lumbar and the first sacral vertebra in spondylolisthesis is awkward for this operation. (2) There are advantages in retaining the loose posterior arch. If satisfactory fusion is obtained its removal is not necessary for the relief of symptoms and its retention provides an island of living bone which facilitates fusion by incorporation of the graft. (3) The fusion should be performed from one vertebra above the defective vertebra to the upper half of the sacrum. In spondylolisthesis of the fifth lumbar vertebra fusion should not be performed higher than the fourth lumbar vertebra. To do so will put undue strain upon the graft which may prevent its solid fusion or may concentrate stresses on too short a segment of mobile lumbar spine and invite osteo arthritis there. (4) Both cortical grafts and cancellous grafts should be used. The cortical grafts should be continuous throughout the field of operation and should be securely fastened to the posterior elements of the vertebrae to act as splints. The cancellous bone should be packed in abundance into all the interstices of the field of operation to ensure rapid osteogenesis. (5) Ample time should be provided for the grafts to fuse solidly enough to resist the stresses of normal use. This cannot be accomplished overnight. It requires not less than 4 months of strict recumbency to give reasonable assurance that the grafts are fused sufficiently to permit return to active life. Even then the field of operation must be protected.

severity of the deformity the physical responsibilities of the patient and the severity of the symptoms

There is much to be said for the treatment of spondylolisthesis by lumbosacral fusion. A successful fusion (solid bridge from the vertebra above to the vertebra below the lesion) will completely relieve the patient of his symptoms and restore him to normal life. Therefore it is the treatment of choice wherever possible but it involves a big operation and is the most difficult of all spinal fusions to accomplish successfully. It demands much time to accomplish solid fusion. There are associated risks and complications (thrombophlebitis, infection, failure to secure bony fusion). Consequently it cannot be undertaken lightly.

Young patients are best suited for treatment of their spondylolisthesis by fusion and the less severe the displacement of the involved vertebra the better will be the result. Young patients with their great capacity for repair are ideal patients for this major operative procedure. Furthermore, because of their youth they can better afford to spend the time necessary for recovery from this operation, and, with their whole active life before them, there is added reason to relieve them of their symptoms and to protect them against progression of the deformity and increase in the severity of their symptoms.

In older people the practicability of lumbosacral fusion becomes increasingly open to question. It is true that a successful lumbosacral fusion will relieve them of all their symptoms, but the magnitude of the operation and its risks and the time which must necessarily be spent in bed and the cost of the procedure must be balanced against the advantages of the relief of symptoms after successful operation. Each case must be judged on its merits.

If operation is contra indicated, much relief can be obtained by a good spinal brace or a corset combined with a supported mattress for the night. The relief will never be complete and it does necessitate the continued use of a cumbersome and restrictive appliance. Operative treatment is better if there is no serious contra indication.

It has been asserted by Gill, Manning and White (1955) that removal of the loose posterior segment of the neural arch alone offers a simple operative procedure that will give the patient quick relief with little loss of time. Some of the pain of spondylolisthesis in some patients is certainly due to the pressure upon a nerve root by the fringe of osteophytes that often surrounds the defect in the neural arch. Removal of the loose posterior fragment and disposal of the osteophytes is likely to relieve the root pressure and the consequent sciatica. It must be remembered, however, that the low back pain which is the most common symptom of spondylolisthesis is not due to root pressure but to instability and is not influenced by removal of the loose posterior arch. Indeed this may increase the instability. It is not sufficiently realized that movement is the last straw that precipitates symptoms in spondylolisthesis, even those symptoms due to root pressure. Immobility and stability are more important than anything else for the relief of symptoms.

The operation of Gill, Manning and White (1955) has not been observed long enough for assessment but it has interesting possibilities. Its simplicity and ease and the short duration of morbidity are its chief merits. These authors have modified their original operation to include if necessary an adequate decompression

of the roots through the intervertebral foramen. Relief of the most severe symptoms may be justification for its use especially in older patients. The residual symptoms may not be severe and can be accepted with some modification of the patient's way of life. It is true also that a certain degree of stability develops in many cases of spondylolisthesis by the growth of a buttress at the antero superior corner of the body of the first sacral vertebra which supports the displaced body of the fifth lumbar vertebra. Increased experience may allow selection of the cases that will obtain most benefit from this operation. One possible objection to the operation is the difficulty in securing fusion of the lumbosacral region should this subsequently be considered necessary. However the experience of Bosworth and his colleagues (1955) perhaps indicates that the difficulty in fusing the lumbosacral region after removal of the neural arch may be more theoretical than practical for they have adopted as their standard procedure for the operative treatment of spondylolisthesis removal of the loose posterior segment of the neural arch plus lumbosacral fusion with an iliac H graft.

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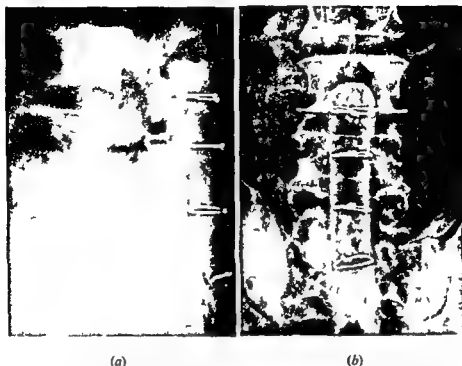


FIG 57 —Lateral (a) and antero posterior (b) radiographs of spondylolisthesis of the fourth lumbar vertebra satisfactorily fused by the principles outlined in the text. The fifth lumbar vertebra is sacralized. Paired tibial grafts were placed on edge and fitted carefully to the laminae and spinous processes from the spine of the third lumbar to that of the second sacral. They were secured to the spinous processes by sutures of stainless steel wire. Cancellous bone was packed into the interstices of the field of operation. Because of the anomalous fifth lumbar vertebra and because the spondylolisthesis involved the fourth lumbar vertebra the length of the spinal fusion is greater than would be necessary for spondylolisthesis of the fifth lumbar vertebra.

with a brace until a year after the operation. (6) Complete stillness after operation can conveniently be achieved by means of a Stryker turning frame. Fig 57 is the radiograph of a satisfactory lumbosacral fusion for spondylolisthesis obtained by the principles outline above.

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(a)

(b)

FIG 57 —Lateral (a) and antero posterior (b) radiographs of spondylolisthesis of the fourth lumbar vertebra satisfactorily fused by the principles outlined in the text. The fifth lumbar vertebra is sacralized. Paired tibial grafts were placed on edge and fitted carefully to the laminae and spinous processes from the spine of the third lumbar to that of the second sacral. They were secured to the spinous processes by sutures of stainless steel wire. Cancellous bone was packed into the interstices of the field of operation. Because of the anomalous fifth lumbar vertebra and because the spondylolisthesis involved the fourth lumbar vertebra the length of the spinal fusion is greater than would be necessary for spondylolisthesis of the fifth lumbar vertebra.

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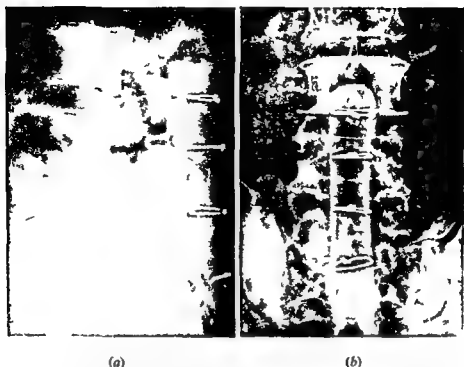


FIG 57 —Lateral (a) and antero posterior (b) radiographs of spondylolisthesis of the fourth lumbar vertebra satisfactorily fused by the principles outlined in the text. The fifth lumbar vertebra is sacralized. Paired tibial grafts were placed on edge and fitted carefully to the laminae and spinous processes from the spine of the third lumbar to that of the second sacral. They were secured to the spinous processes by sutures of stainless steel wire. Cancellous bone was packed into the interstices of the field of operation. Because of the anomalous fifth lumbar vertebra and because the spondylolisthesis involved the fourth lumbar vertebra the length of the spinal fusion is greater than would be necessary for spondylolisthesis of the fifth lumbar vertebra.

with a brace until a year after the operation. (6) Complete stillness after operation can conveniently be achieved by means of a Stryker turning frame. Fig 57 is the radiograph of a satisfactory lumbosacral fusion for spondylolisthesis obtained by the principles outlined above.

ACKNOWLEDGEMENTS

The generous assistance of my colleagues has made possible the compilation of the material to illustrate this chapter. Special thanks are due to Dr D Wollin of the Department of Radiology of the Toronto General Hospital, Dr John Munn of the Department of Radiology of the Hospital for Sick Children, Professor J W A Duckworth, Department of Anatomy, University of Toronto, and many other friends for radiographs of their patients.

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CHAPTER 3

STRUCTURAL SCLIOSIS

J I P JAMES

Scoliosis is the term applied to all lateral curves of the spine in a structural scoliosis alone is there a fixed rotation

A structural scoliosis is easily recognized. Examination of the back of the standing patient will show a lateral curve of the spine with compensatory curves above and below the main or primary curve. On forward flexion the compensatory curves straighten out, but in the primary curve or curves the rotation and lateral curvatures persist (Fig 58)

The site of the primary curve is determined by clinical examination and may involve any part of the vertebral column. There is usually one primary curve with a compensatory curve above and below it (3 curves in all). In congenital scoliosis the primary curve may involve the lumbosacral region and as there is then no room for a compensatory curve below there will be a single compensatory curve above (2 curves in all). Sometimes there are 2 primary curves that is 2 curves in which rotation persists on forward flexion. These curves always adjoin and are opposite in direction. Compensatory curves will be present above and below the 2 primary curves (4 curves in all).

In addition to the site and number of the primary curves the aetiology should be established if possible as a guide to prognosis and treatment.

AETIOLOGICAL CLASSIFICATION

Cobb (1948) has suggested a useful aetiological classification and this has been slightly modified

Structural scoliosis

- 1 Idiopathic
- 2 Osteopathic
 - a Congenital
 - b Osteo-chondrodystrophy
- 3 Neuropathic
 - a Congenital
 - b After poliomyelitis
 - c Neurofibromatosis
 - d Other neuropathies
- 4 Myopathic
 - a Congenital
 - b Muscular dystrophy
- 5 Thoracogenic
 - a After empyema
 - b After pneumonectomy or thoracoplasty

Idiopathic scoliosis

This is not associated with any other pathological condition and is therefore diagnosed by a process of exclusion. Confirmation is obtained from the site, length and mobility of the curve and is discussed later.

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Neurofibromatosis is complicated by scoliosis in some 10 per cent of cases. Although the full picture of this disease may be present—with café au lait patches, numerous cutaneous neurofibromas and so on—scoliosis is more often seen with only minor stigmata. A few café au lait patches should always arouse suspicion.

PROGNOSIS

The prognosis in the various types of scoliosis will be discussed at length. At present the principal method of treatment of a structural scoliosis is by correction and fusion of the primary curve. This is somewhat disappointing if performed at the end of growth when the deformity is complete and the results are improved by earlier surgical intervention. The indications are necessarily based on the expected development of the curve. Detailed knowledge therefore of the various aetiologies and curve patterns is essential if operation is to be undertaken at the right time during development.

IDIOPATHIC SCOLIOSIS

In idiopathic scoliosis the prognosis is largely determined by the site of the primary curve (Ponseti and Friedmann 1950; James 1954). There is however one other factor—the age at which the curve begins. As would seem obvious, the earlier scoliosis appears, the more severe is the curvature at the conclusion of growth. But the development of each curve is determined more by its site in the vertebral column than by the age of onset. In the same segment of the spine the age at which the curve began provides an added refinement of prognosis.

Scoliosis is progressive only during skeletal growth. After skeletal maturation an increase need not be feared, although in paralytic scoliosis a small increase may exceptionally occur. Spinal skeletal maturation is indicated by the growth of the iliac apophysis, which appears anteriorly and extends backwards. Risser and Ferguson (1936) noted that spinal growth stopped when the iliac apophysis reached and turned down towards the posterior superior iliac spine (Fig. 59). Mature curves are those in which the iliac apophysis has completed this stage.

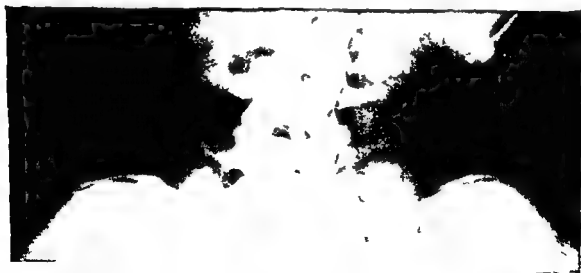


FIG. 59—The iliac apophyses have reached the posterior superior spines and have just begun to turn down towards them. This coincides with the cessation of vertebral growth. (By courtesy of the Editor of *J. Bone Jt. Surg.*)



FIG 58—In the erect and bending positions it can be seen that there is one curve with fixed rotation and the cervical and lumbar compensatory curves to the right do not have rotation on forward flexion

Congenital scoliosis due to vertebral abnormalities is most easily recognized by radiography but there may also be other congenital abnormalities, subcutaneous angiomas spider naevi hairy patches skin dimples and fatty tumours are common and if a spina bifida is present there may be motor paralysis and impairment of sensation in the lower limbs. Absent ribs may be betrayed by a visible sulcus in the chest wall and sometimes by absence of one pectoralis major and breast. A short wide neck and a low hair line often betoken a high thoracic congenital curve. Pelvic obliquity occurs in nearly all severe lumbosacral congenital curves.

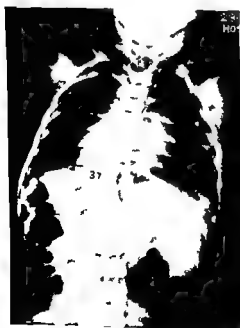
Paralytic scoliosis may be accompanied by obvious paralysis or paresis may be apparent only after complete muscle examination and charting which consequently is always necessary. In some paralytic scolioses with little or no limb involvement the original attack of poliomyelitis may have been undiagnosed because of the atypical distribution of paralysis. Severe involvement of the intercostals is often mistaken for pneumonia.

Neurological conditions such as syringomyelia Friedreich's ataxia and less commonly a Charcot Marie Tooth neuropathy may cause scoliosis. A thorough neurological examination must therefore be undertaken particularly if the curve pattern is atypical. A common neuropathic pattern of scoliosis is a double primary curvature in which the primary curves do not balance and gross listing or decompensation exists.

developed before the age of 10 years in about one third of these children and quite often before the age of 4-6 years (Fig 60). Occasionally progression is slow but it is extremely rare to see a curve of less than 70 degrees at the age of 10. Almost invariably by the end of growth there is a curve of at least 70-100 degrees. Amongst all the scolioses there is no curvature with a consistently worse prognosis or that so commonly attains 100 degrees. It is in these severely deformed and crippled children that dwarfing occurs.

There are two exceptions to the bad prognosis. In some children who present this type of curve from the age of 2 or 3, the curvature increases very slowly and at from 4-6 years of age there appears a lumbar curve with fixed rotation. It is then apparent that curves originally diagnosed as infantile thoracic have become converted to infantile double primary curves with a much improved prognosis. The other exception is infantile resolving structural scoliosis.

FIG. 60—(a) Infantile idiopathic left thoracic scoliosis. (b) The initial curve of 37 degrees showing no congenital anomalies. (c) By the age of 6 years it had reached 118 degrees.



Measurement of the angle of curvature is essential and the method of Cobb (1948) is used. The upper and lower vertebrae of a primary curve show no rotation of the spinous process and the disc above and below them is not wedged. From the upper and lower borders of these vertebrae respectively perpendiculars are erected the angle at which they meet is the angle of curvature.

Mild curves are defined radiographically as those between 0-69 degrees severe curves 70-99 degrees and very severe curves 100 degrees or more.

Cervico thoracic scoliosis

Ponseti and Friedmann (1950) described 5 cases in some 400 patients with idiopathic scoliosis. The author has not found a case among almost 2 000 patients with scoliosis of various aetiologies. Ponseti reported a good prognosis.

Thoracic scoliosis

Many cases of idiopathic scoliosis occur in the thoracic spine and they form the most important group. In order to determine the prognosis more accurately the cases are divided according to the age of onset of the curve.

Infantile	age of onset 0-3 years
Juvenile	age of onset 4-9 years
Adolescent	age of onset 10 years to the end of growth

Infantile idiopathic thoracic scoliosis

Scoliosis in infants is not uncommon and arises from most of the conditions already discussed. When however the scoliosis of known aetiology are excluded there is a residual group to which the term idiopathic has been applied (James 1951). The group is homogenous and the curvature important.

The sex incidence is equal and for some unknown reason 90 per cent of these curves are to the left. Adolescent thoracic idiopathic scoliosis has by contrast an incidence of almost 90 per cent in girls and 90 per cent of the primary curves are to the right.

The age at which this curve is first seen varies but is usually between 6 and 18 months. Among the 120 infants seen with this type of scoliosis in only 2 was it noticed at birth. Although the infants are apparently otherwise normal at first, they are later often undersized although they have no endocrine abnormalities. Cranial moulding and facial hemiatrophy also occur.

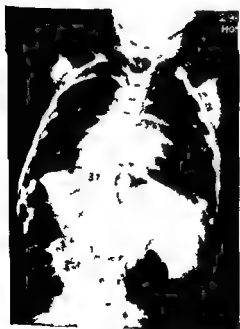
The lateral curvature may be very small but there is necessarily if it is to be included in the group of structural scolioses a rib rotation. In the early stages this is not easily discerned except on forward flexion. The radiograph shows a lateral curvature with compensatory curves above and below and with visible rotation of the vertebral bodies of the primary curve. The diagnosis of an idiopathic curve can only be finally established by a radiograph showing no vertebral anomalies. If a patient is seen later in life when there is marked secondary wedging and other changes it will be impossible to establish the true aetiology without early films. Vertebral anomalies can be small and difficult to see. In the large number of children who have now been operated upon for infantile idiopathic thoracic scoliosis no vertebral laminal anomalies have been found a confirmation that the scoliosis is not congenital.

Usually the curvature is rapidly progressive. Curves of more than 100 degrees

developed before the age of 10 years in about one third of these children and quite often before the age of 4-6 years (Fig 60). Occasionally progression is slow but it is extremely rare to see a curve of less than 70 degrees at the age of 10. Almost invariably by the end of growth there is a curve of at least 70-100 degrees. Amongst all the scolioses there is no curvature with a consistently worse prognosis or that so commonly attains 100 degrees. It is in these severely deformed and crippled children that dwarfing occurs.

There are two exceptions to the bad prognosis. In some children who present this type of curve from the age of 2 or 3, the curvature increases very slowly and at from 4-6 years of age there appears a lumbar curve with fixed rotation. It is then apparent that curves originally diagnosed as infantile thoracic have become converted to infantile double primary curves with a much improved prognosis. The other exception is infantile resolving structural scoliosis.

FIG. 60—(a) Infantile idiopathic left thoracic scoliosis. (b) The initial curve of 37 degrees showing no congenital anomalies. (c) By the age of 6 years it had reached 118 degrees.



Infantile resolving structural scoliosis

Amongst 120 infants with idiopathic structural scoliosis there were 20 in whom the curve disappeared. Although they had a structural curve this was never large and was often without compensatory curves. Sometimes the radiograph showed that the lateral curvature was minimal but that rotation of the vertebral bodies was pronounced (Fig. 61).

Two radiographic features enable us to expect the spontaneous disappearance of a curve: first a small primary curve without compensatory curves, or secondly marked rotation and little lateral curvature. Clearly the progressive type may also be seen at an early stage without compensatory curves and this particular radiographic finding is less certain than the disproportionate vertebral rotation. The



(a)



(b)

FIG. 61—Infantile resolving structural scoliosis. (a) Radiograph showing almost no lateral curvature but marked rotation of the vertebral bodies. (b) A year later the spine had become normal.

interesting feature of this curve is that undoubtedly there is a structural scoliosis and yet for reasons unknown to us the curve disappears. Scott (1956) named this group the resolving structural scolioses of infancy. Unfortunately there is as yet no certain way of distinguishing the progressive from the resolving structural scoliosis in the earlier stages, but observation over a few months establishes which pattern is present.

Denis Browne (1956) regards infantile idiopathic scoliosis as a congenital postural curve. He considers that abnormal intra uterine pressure leads to scoliosis *in utero*. Although it is exceptional for scoliosis to be present at birth—2 cases out

of 120 in this series—he believes that the curve can be demonstrated by an impairment of lateral flexion in one direction immediately after birth. His nomenclature is open to criticism because if the condition were truly congenital, one would expect it to be more obvious at birth and also the term postural should not be applied to curves in which true structural changes occur. Denis Browne believes that this scoliosis can be cured by early splintage but others disagree. It is probable that the true incidence of resolving structural scoliosis is much greater than in the present series which comprises cases referred to a scoliosis clinic. Here the more serious and later developed curves tend to be seen, whereas in paediatric units the earlier and resolving types are commonly found.

Scott (1956) has referred to the frequency of postural scoliosis of infants. This disappears but being without fixed rotation or fixed curvature, it is not relevant to this chapter.

Juvenile idiopathic thoracic scoliosis

Thoracic scoliosis is occasionally found amongst children of 4–10 years of age the incidence of right and left curves being equal. The earlier onset makes the prognosis somewhat worse than in the adolescent type from which otherwise it does not differ. Some 80 per cent develop a curve of at least 70 degrees by the end of skeletal maturation.

Adolescent idiopathic thoracic scoliosis

The typical features of a thoracic scoliosis are well illustrated in adolescents. There is marked dropping of the shoulder on the side of the concavity and rotation of the ribs posteriorly on the convexity and forwards on the concavity. The iliac crest on the side of the concavity becomes prominent and a very unsightly deformity occurs (Fig. 62). This type of idiopathic thoracic scoliosis is most often right sided and occurs in girls—the classical text book scoliosis. The curvature may increase rapidly and within 2 years a curve of more than 100 degrees may develop.

There were 66 patients in this group who had reached skeletal maturation. In 59 per cent there were curves of more than 70 degrees and the range was from 24 to 151 degrees (Fig. 66).

Thoraco lumbar idiopathic scoliosis

In general the lower a primary curve is in the spine the better the prognosis. In the thoraco lumbar scoliosis the apex of which involves the thoracic eleventh or twelfth vertebrae is mid way in prognosis between the thoracic and the lumbar curves. It rarely develops before the age of 10 years. It is most common in girls and is most often to the right. Although a dropped shoulder and particularly a prominent hip are obvious features the rib rotation since some of the involved vertebrae have no ribs is less obvious and therefore the appearance even for the same degree of curvature is less unsightly than in thoracic scoliosis (Fig. 63). Thoraco lumbar scoliosis is somewhat rare. Of 26 patients with this pattern who had reached skeletal maturation there were 18 (69 per cent) with mild curves and 8 (31 per cent) with severe curves. No curves exceeded 100 degrees.



FIG 63—Adolescent idiopathic thoracic scoliosis. (a) The classical right sided curve of the adolescent girl. (b) A curve of 116 degrees developed between the ages of 14 and 16 years.

Idiopathic lumbar scoliosis

Lumbar scoliosis is a common curve rarely seen before the age of 10 years. It affects girls much more often than boys but shows no preference for right or left. Although prominence of the hip is usual, the curve is small and there is no dropping of the shoulder and little rib rotation (Fig 64). Consequently the cosmetic effect is limited and many patients attain adult life before the curve is discovered. Amongst 79 patients with mature curves 72 (91 per cent) had curves of less than 70 degrees and many of these were less than 30 degrees. Only 7 patients (9 per cent) developed curves of more than 70 degrees and none had a very severe curve.

Although in terms of curvature and appearance lumbar idiopathic scoliosis has an excellent prognosis it does cause one serious problem in later life. It is common to find a mild lumbar scoliosis in a woman complaining of backache. A radiograph shows sclerosis and osteoarthritic degeneration of the posterior interarticular joints confirmed clinically by stiffness. The pain is intractable and may be referred pain down the back of the thighs to the knee but straight leg raising is not limited. Late backache develops in many patients with idiopathic lumbar scoliosis although often delayed until the age of 50 or 60 years of age. Occasionally the curvature is increased by postmenopausal or senile osteoporosis.



FIG 64 —Thoraco lumbar idiopathic scoliosis (a) Note the very prominent iliac crest characteristic of this particular scoliosis (b) The apex is at the eleventh thoracic vertebra

Combined thoracic and lumbar idiopathic scoliosis

The idiopathic curves so far discussed have had one primary curve. There is a common pattern, however, presenting 2 primary curves, one thoracic, the other lumbar. Usually they are almost equally severe, but sometimes one may predominate. The classical pattern has certain important features, the curves being equal, the rotations compensate one for the other. There is little or no shoulder dropping and the hips are not very prominent. This curve therefore is never very apparent and at its worst is likely to disfigure by the shortness of the trunk rather than by the usual stigmata of scoliosis (Fig 65). The age at which the curve begins is variable, usually after 10 years, often between 4 and 8 years, and sometimes in infants as noted already.

Many double primary curves are slow to progress and like the lumbar remain mild and hardly noticeable. Among 54 mature cases there were 38 patients who had curves of less than 70 degrees (70 per cent), 13 with severe curves (24 per cent) and 3 with very severe curves (6 per cent) in which both primary curves were more than 100 degrees. These latter curves all started before 10 years of age. It is usual for those with a late onset to have right thoracic, left lumbar curves, those with an early onset often have left thoracic curves and there is an increased incidence in males.

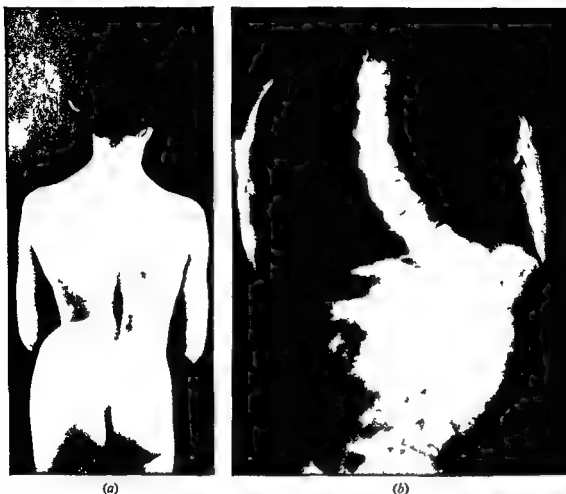


FIG 64 —(a) and (b) Idiopathic lumbar scoliosis. A curve with a good prognosis both in appearance and the degree of curvature to be expected

Intermediate idiopathic scoliosis

Ponseti described a large number of cases of combined double primary idiopathic scoliosis, a higher percentage than in the present series. However, his diagnostic criteria for a double primary curve were largely radiological, whereas this series has been based predominantly on clinical examination, with the radiographic appearances being regarded as secondary. By clinical and radiographic examination, there is a small group that is intermediate between those in which there is a single primary curve and those with two curves.

If we diagnose a primary curve by the persistence of rotation on forward flexion, then all compensatory curve rotations should disappear on forward flexion. This intermediate group described by Ponseti consists of a small number of patients with primary thoracic curves in whom a minimal persistent lumbar rotation is apparent on forward flexion. This rotation is small radiographically and clinically, and one could not regard the lumbar curves as second primary curves, but equally it is impossible to ignore this rotation and classify them as typical compensatory curves. From the point of view of correction and fusion, the lumbar rotation may be disregarded; in other words, the lumbar curve is treated as compensatory, and it will not increase after fusion of the thoracic curve alone.



FIG 65—Combined thoracic and lumbar idiopathic scoliosis. Note the good appearance (a) and the two primary curves of almost the same amount 68 and 66 degrees (b) (B's courtesy of the Editor of *J Bone Jt Surg*)

Summary of prognosis in idiopathic scoliosis

It is possible now to summarize the prognosis of these idiopathic groups. The age at which the curve starts is important but is clearly subordinate to the site of the primary curve. As we go down the spine the prognosis improves so that the prognosis in terms of degrees of curvature of a thoracic curve is notably worse than that of a lumbar curve. Fig 66 shows this in tabulated form. The curves from which this analysis is made were all mature.

Although the thoracic curve has a poor prognosis in degrees of curvature and a lumbar a good prognosis it is not only in degrees of curvature that the prognosis differs. The appearance of the patients with curves of the same degree but in different situations is shown in Fig 67. In each case there is a primary curve of 70 degrees. The lumbar curve on the left shows minimal deformity particularly if the patient is clothed. A thoraco lumbar curve is more apparent while a thoracic curve produces a gross deformity. A combined curve of this degree is barely noticeable.

CONGENITAL SCLIOSES

Congenital scolioses are associated with demonstrable vertebral anomalies. These anomalies are very varied. There may be one or more hemivertebrae, congenital absence of discs or fusion of vertebral bodies, absence of ribs, rib fusions or a bizarre collection of anomalous vertebrae extending through the whole or most of the vertebral column.

STRUCTURAL SCLIOSIS

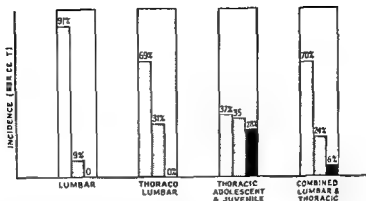


FIG 66 —Idiopathic scoliosis prognosis according to pattern of curvature. Incidence of mild severe and very severe curves in each of the four major curve patterns. Mild curves are represented by light shading, severe curves by dark shading, and very severe curves in black. Compare with Fig 67. (By courtesy of the Editor of *J Bone Jt Surg*)

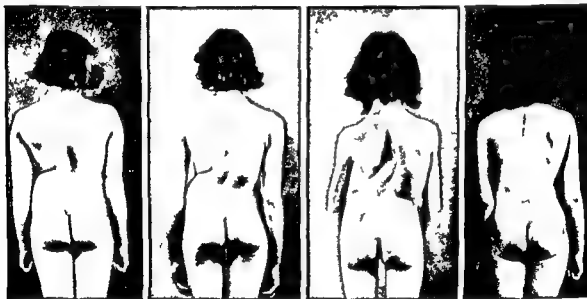


FIG 67 — A photographic comparison of the same four types of idiopathic scoliosis: lumbar, thoraco-lumbar, thoracic, and double primary. Each girl has 70 degrees of scoliosis. (By courtesy of the Editor of *J Bone Jt Surg*)

In general the isolated vertebral anomalies have a good prognosis. It is not uncommon to detect a congenital scoliosis late in childhood, often this does not develop into more than a minimal curve, and most do not progress and require no treatment.

There are, however, cases of congenital scoliosis, particularly in the thoracic region and also those involving a long vertebral segment, in which gross scoliosis develops. There are no more severe curves than those sometimes seen in the congenital scolioses that produce asymmetrical growth and curvature (Fig 68). Unfortunately it is not easy at the beginning to recognize which type is present, although the curve that is likely to produce a gross deformity later will usually start advancing from early childhood.



FIG 68 —Severe congenital scoliosis. There are numerous congenital anomalies of the vertebrae and a right angled deformity at the apex with vertebral fusion.



STRUCTURAL SCOLIOSIS

There is one type of congenital scoliosis in which several anomalous upper thoracic vertebrae show clinically no fixed deformity but there develops below a severe thoracic primary curve in which there are no anomalous vertebrae that is the congenital anomalies lie in the compensatory curve above the primary curve (Fig 69). Although there is much to suggest that the congenital anomalies are incidental and that the patient is developing an idiopathic thoracic curve with whose behaviour it accords the curve must necessarily be classed as congenital. The prognosis of this particular type is as bad as for the thoracic idiopathic curves.

Congenital scolioses more than any others lead to paraplegia usually in the later years of growth. Congenital scoliosis therefore must remain under observation until the end of growth in order to detect early paraplegia this must be done even if the curvature remains mild and unchanging.

PARALYTIC SCOLIOSIS

There have been few attempts to assess the prognosis of paralytic scoliosis. Colonna and Vom Saal (1941) tried to correlate muscle paralysis with the development of curves and attempted to distinguish the various patterns. However, their work was particularly related to correlating muscle paralysis and curvature.

Recently James (1956) reviewed 193 patients with paralytic scoliosis. Curve patterns resembled those found in idiopathic scoliosis, but some patterns in paralytic scoliosis were not found in the idiopathic group. In paralytic scoliosis the curves tend to be longer and remain more mobile but rib rotation is more sharply angular and razor edge rotation is common.

In order to correlate the paralysis with the type of curve it is important to recognize the various patterns by the site of the primary curve. This bears some relationship to prognosis but the prognosis of paralytic scoliosis in contrast with that of idiopathic scoliosis is not determined by the anatomical site so exclusively. As will be shown later prognosis is largely determined by the muscle imbalance and by the age at which poliomyelitis occurred. The number of mature patients with paralytic scoliosis in our series is limited and in some patterns the number was too small to draw significant conclusions. In the first place however, a review of the patterns in paralytic scoliosis with their prognosis will be presented.

Paralytic curve patterns

High thoracic	39
Thoracic	69
Thoraco lumbar	47
Lumbar	17
Combined thoracic and lumbar	13
Telescopic spine	8
Total	193

High thoracic paralytic scoliosis

This pattern in which the curve began at the seventh cervical vertebra (1 case), the first thoracic vertebra (31 cases) or second thoracic (7 cases) does not exist in idiopathic scoliosis.

Although separation of the high thoracic pattern from the ordinary thoracic curve may appear scarcely justifiable there are two reasons for doing so. First

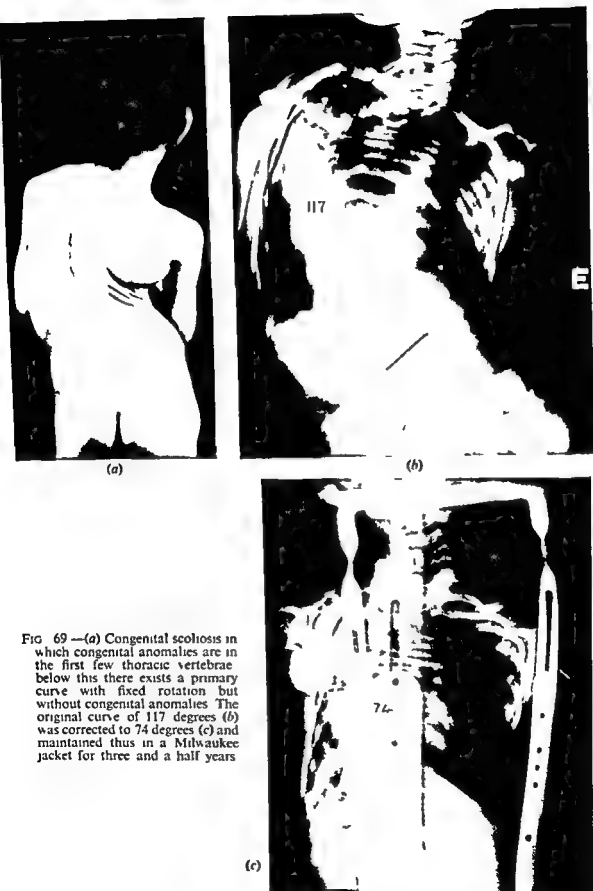


FIG. 69 —(a) Congenital scoliosis in which congenital anomalies are in the first few thoracic vertebrae below this there exists a primary curve with fixed rotation but without congenital anomalies. The original curve of 117 degrees (b) was corrected to 74 degrees (c) and maintained thus in a Milwaukee jacket for three and a half years

its prognosis is worse than the lower thoracic curve and secondly the clinical appearance of the patient is characteristic and differs from that in other paralytic scolioses. The first and second ribs on the convexity are elevated and rotated backwards thus producing elevation of the shoulder neck line and the characteristic appearance. As the primary curve develops, there is an increasing compensatory cervical curve which displaces the head sideways and produces a very ugly deformity. The main cosmetic defect is caused by the rib rotation and this is not affected by surgical correction of the scoliosis (Fig 70). The only good results from this pattern can be those in which operation is undertaken before severe deformity occurs.

Twenty of the 39 patients had reached skeletal maturation, after which a paralytic scoliosis rarely increases appreciably. A corollary is that structural scoliosis does not develop in a patient who contracts poliomyelitis as an adult. Among the 20 patients there were 4 with mild scoliosis, 1 with severe scoliosis and 15 with very severe curves (more than 100 degrees) one of whom had a curve of 155 degrees.

Although the remaining 19 patients are not mature there are already 4 curves of more than 100 degrees and 9 with curves of more than 70 degrees. Of 6 whose curves remain mild, 4 are less than 10 years of age. In 11 of these 19 immature patients correction and fusion have already been carried out because of the rapid progression of their curves.

Thoracic paralytic scoliosis

This curve proved to be one of the commonest of the paralytic scolioses and its prognosis relatively poor, not only because of the degree of curvature but because, as with idiopathic scoliosis, it produces a marked change in the appearance. Although the incidence between the sexes is about equal there is an unexplained predominance of curves to the right (54 compared with 15 to the left). The length of these curves was characteristically longer than in idiopathic curves and involved as many as 13 vertebrae (Fig 71).

Thirty patients had completed growth, 8 of whom had mild curves, 10 severe and 12 very severe. This is better than in the high thoracic group. In the patients still immature there were only 9 curves of more than 100 degrees, most of these patients had already undergone fusion.

Thoraco lumbar paralytic scoliosis

Paralytic scoliosis may produce extremely long curves which have been termed C curves and were said to involve the whole vertebral column. However close study of the radiograph shows that there are upper and lower compensatory curves although these may be very short. In paralytic scoliosis in general and in thoraco lumbar in particular there is often a gross list or decompensation. This is partly because of the length of the curve and the difficulty of compensation in the short remaining length of vertebral column and also because of the muscle weakness with inability to hold the erect position (Fig 72).

The term thoraco lumbar was restricted to curves having their apex at the eleventh or twelfth thoracic vertebrae. There were 47 patients in whom the sexes were of equal number and there were twice as many curves to the right as to the left. Of these 47 patients only 18 had completed spinal growth and it is therefore difficult to have complete data concerning prognosis. There were, however



(a)



(b)

FIG 70—High thoracic paralytic scoliosis (a) and (b) Note the elevated shoulder neck line due to the rotation of the first and second ribs (c) The curve extends between the first and sixth thoracic vertebrae the ribs on the apex droop the only paralysis existing was that of the apical intercostals on the convexity



(c)

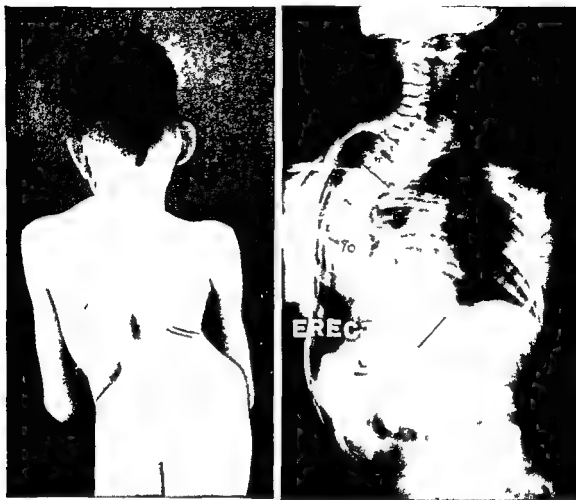


FIG 71 —Thoracic paralytic scoliosis - Note the collapsed ribs opposite the apex on the convexity

7 patients with mild curves 4 with severe and 7 with very severe curves (more than 100 degrees)

Lumbar paralytic scoliosis

Unlike idiopathic curves these were uncommon there being only 17 patients 12 of whom had completed growth but of these 2 were mild 7 were severe and 3 were very severe The prognosis is like that in other patterns of paralytic scoliosis but compares most unfavourably with its idiopathic counterpart (Fig 73)

Lumbar scoliosis like thoraco lumbar paralytic scoliosis tended to be of two types the first due to a muscle imbalance and the second to a symmetrical severe muscle weakness in which gravity caused collapse at first postural later structural Several patients showed a flail or telescopic collapse on assuming the erect position but the curve disappeared on lying down The transition from a postural paralytic curve to a structural curve is interesting

These collapsing spines were uncommon and tended to develop lumbar or thoraco lumbar curves as time passed However in a child suffering poliomyelitis



FIG 72 —Thoraco-lumbar paralytic scoliosis. This boy's curve has continued to progress. He has considerable weakness of the trunk muscles and is now ready for correction and fusion.

late in growth there might never be a structural curve but only a marked collapse on assuming the erect position.

A small number of patients showed two primary curves. These curves were less balanced than in idiopathic scoliosis and did not carry the same good prognosis. It is likely that these two primary curves developed as a result of fortuitous asymmetry of paralysis on one side above and on the other side below. They are included for completeness: both curves must be fused if correction is undertaken.

Summary of prognosis in the paralytic scolioses

The prognosis of the various types of paralytic scoliosis has been briefly discussed and a comparison of the 4 major groups is tabulated for comparison. The siting of the primary curve is obviously much less significant than in idiopathic scoliosis (Fig 74).

The most obvious prognostic factor is the age at which the disease first affected the child. Although curvature of the spine is often noticed within a few weeks or months of the initial disease it is sometimes delayed, particularly in the high thoracic paralytic scoliosis. In one patient there was an interval of 14 years between the onset of the disease and of curvature. However, as will be seen from



Fig 73 —Lumbar paralytic scoliosis There was a weakness of the lateral abdominal flexors on the convexity and a general symmetrical trunk weakness

Fig 75 most paralytic scolioses developed within 2 years of the disease and thereafter there was a sporadic development of curvatures in the whole group

Children who developed poliomyelitis before the age of 5 years usually matured with curves of more than 100 degrees whereas those children who developed poliomyelitis after the age of 10 years mostly ended with curves of less than 70 degrees (Fig 76) These curves were about equally distributed between the various patterns and the only obvious difference since the muscle paralyses were much the same was the age of onset of the original disease It is clear therefore that the longer the imbalance of muscle has to affect vertebral growth the more distortion there will be and the worse the final result

Muscle paralysis in paralytic scoliosis

The correlation of curvature with the particular muscles paralysed is complex and few clinics have sufficient material The only serious attempt was made by Colonna and Vom Saal who besides demonstrating a relationship between paralysis of the trunk muscles and spinal deformity considered the limb muscles important the strongest muscles in the upper limbs drawing the spine into a convexity on the stronger side

Mayer (1936) stressed the importance of the lateral flexors in causing pelvic obliquity and lumbar scoliosis Colonna and Vom Saal agreed with him and

PROGNOSIS

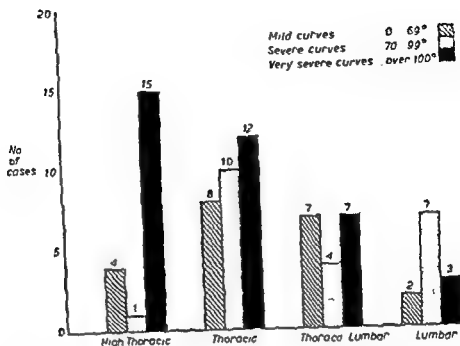


FIG 74—Prognosis of paralytic scoliosis patterns. A large number of mature curves was not available and the figures are therefore rather small for accuracy.

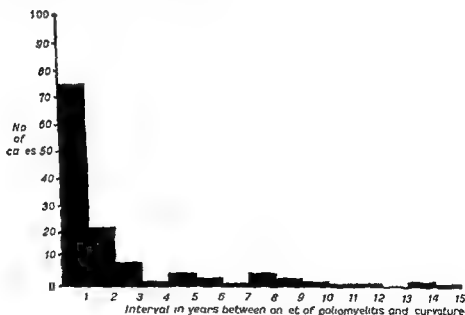


FIG 75—Interval between the onset of poliomyelitis and the onset of curvature taken from a series of 132 patients. It is clear from this that most paralytic curves appear in the first year but there is a steady appearance of occasional curvature after many years.

in this present series this finding is confirmed. Otherwise the findings of Colonna and Vom Saal (1941) have not been confirmed.

To find the significant muscle weakness causing paralytic scoliosis the muscle paralyses of 193 patients were tabulated. The scolioses were divided into the

STRUCTURAL SCLIOSIS

patterns previously described and within each pattern the muscles paralysed were recorded. The significant group of muscles could thus be discovered. It was difficult to show any correlation between limb muscle paralysis and the development of a paralytic scoliosis. This finding it is believed is reliable because the charting of limb muscles is relatively accurate. The importance of the trunk muscles will be discussed but it should be stressed that the accurate charting of trunk muscles is not easy and certainly not accurate. However, asymmetry of trunk muscles is easy to demonstrate and it is believed that the following impressions are significant

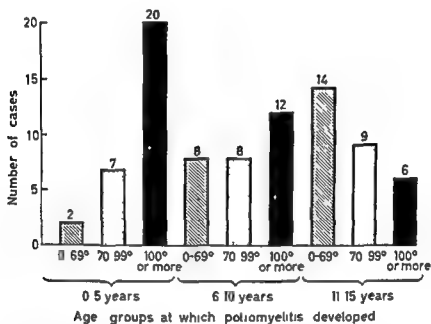


FIG. 76 —Relationship of final curvature to age at original attack of poliomyelitis. It is obvious that the very severe curves (black) are related to the age at onset of the disease. (By courtesy of the Editor of *J. Bone Jt. Surg.*)

High thoracic scoliosis and lumbar scoliosis will be discussed in detail because these show the most clear cut relationship of paralysed muscles to curvature and the general position found in the other curves will be mentioned.

Muscle paralysis in high thoracic paralytic scoliosis

The distal arm muscles played no part and these are not listed here. The arm/trunk muscles as a group were compared on each side.

High thoracic curves—arm to trunk muscles

normal both sides	15
weak on the convexity	10
weak on the concavity	6
symmetrically weak	8

Total 39

PROGNOSIS

The latissimus dorsi which was particularly referred to by Colonna and Vom Saal was found to have the following relationship

Latissimus dorsi on both sides grade 4 to normal	20
weak on the convexity	7
weak on the concavity	6
equally weak or no record	6
Total	39

There were individual asymmetrical paralyses of certain muscles which were difficult to correlate as their number was small. The main findings were as follows

Trapezius weak on the convexity	5
weak on the concavity	4
Serratus anterior weak on the convexity	5
weak on the concavity	3
Pectoralis major weak on the convexity	5
weak on the concavity	4
Rhomboids weak on the convexity	8
weak on the concavity	4

This group reveals no significant relationship between the side of shoulder paralysis and the side of curvature and many patients had no weakness of either side. A slight predominance of weakness on the convexity (the opposite of that noted by Colonna and Vom Saal) is probably because weakness of the trunk muscles on one side accompanies weakness of the limb of the same side; this as would be expected from the nature of the affection of the anterior horn cells.

Comparison of the trunk muscles showed the following

<i>High thoracic curves—intercostals</i>	
weakness towards the convexity	29
weakness towards the concavity	0
symmetrically weak (less than 2) or totally absent	5
inadequate records	5
Total	39

<i>High thoracic curves—erector spinae</i>	
grade 4 to normal	32
less than 4	6
no record	1
Total	39

<i>High thoracic curves—lateral abdominal flexors</i>	
symmetrical grade 4 to normal	26
weak on the convex side	4
weak on the concave side	5
no records or symmetrically weak less than grade 4	4
Total	39

STRUCTURAL SCOLIOSIS

<i>High thoracic curves—abdominal muscles</i>	
grade 4-5 symmetrical	18
less than grade 4 symmetrical (negative Beevor's sign)	16
asymmetrical weak towards convexity	2
asymmetrical weak towards concavity	2
no record	1
	<hr/>
Total	39

From this review of the trunk muscle paralyses there seemed to emerge only one that was significant intercostal weakness on the convexity. Otherwise there was for instance no correlation between abdominal muscles paralysed on the convexity or on the concavity. Although it has not before been suggested that intercostal muscles are related to paralytic scoliosis and although there are few muscles more difficult to chart accurately it is believed that this finding is significant. In 6 patients only intercostal paralysis could be found all other muscles being normal.



FIG 77—Rib descent. The remarkable rib collapse seen in intercostal paralysis causing paralytic scoliosis is particularly well illustrated in this radiograph.

these patients had curvatures ranging from 77 to 127 degrees. These are severe curves and suggest that the intercostal paralysis played an important part.

A further and significant point in the relationship of intercostal paralysis to the development of thoracic scoliosis was the phenomenon of rib collapse or descent. With almost no exceptions ribs were found to have descended and crowded together on the convexity and this appearance of rib collapse or descent proved a reliable radiographic indication of the aetiology of the thoracic scoliosis. It is believed that this rib collapse (Fig 77) is caused by the weakness of the intercostals which no longer hold up the ribs and it was not found in patients without

PROGNOSIS

intercostal paralysis. A surprising finding in this group was the number of patients with normal erector spinae muscles.

To test the impressions gained from patients with paralysis of trunk muscles and scoliosis a survey was carried out on 280 patients suffering from paralysis but without scoliosis. Thirty-eight patients who had markedly asymmetrical arm paralysis for an average duration of 7 years (and never less than 4 years) showed no thoracic scoliosis. In most the arm paralysis was severe, in some it followed injections—a sequence that often causes severe poliomyelitis do so within this time (Fig. 75) and the absence of scoliosis is unlikely to be fortuitous.

Lumbar paralytic scoliosis

The number of patients in this group is small. The muscle paralysis found in 17 patients was as follows:

<i>Erector spinae</i>	
grade 4 to normal	6
less than 4 symmetrical	11
	<hr/>
	17
<i>Lateral abdominal flexors</i>	
grade 4 to normal	3
symmetrically weak (less than 4)	2
asymmetrically weak on the convexity	12
asymmetrically weak on the concavity	0
	<hr/>
Total	17
<i>Anterior abdominals</i>	
grade 4 to normal	3
symmetrically weak (less than 4)	11
asymmetrically weak on the convexity	2
asymmetrically weak on the concavity	1
	<hr/>
	17

There are two significant findings: Although no patient had weakness of the lateral abdominal flexors on the concavity, 12 had weak lateral flexors on the convexity. In all these 12 patients there was associated pelvic obliquity as described by Mayer. The pelvic obliquity and the lumbar scoliosis could be correlated with the marked asymmetry of the lateral flexors so that the strong lateral flexors pulled the rib and pelvis together on the strong side, producing a convexity on the weak side.

A small number of patients presented a severe weakness of the erector spinae, the abdominal muscles and also a symmetrical weakness of the lateral flexors—not an imbalance but a severe symmetrical paralysis. Although the final result was a lumbar scoliosis the mechanism differed from the asymmetrical lateral flexor paralysis just described. In severe balanced paralysis the body weight collapsed the spine and ultimately produced a structural lumbar curve with rotation not to be differentiated from that caused by asymmetrical muscles. Both factors frequently operate.

As with the upper limb, no correlation could be demonstrated with weakness of the muscles of the lower limb. There was no relationship to contracture of the tensor fasciae latae nor true shortness of a limb.

Although the two groups of paralytic scoliosis discussed here in detail show a fairly clear cut correlation with muscle paralysis, thoracic scoliosis and thoraco lumbar scoliosis were not so clearly related to asymmetrical paralysis. In most however it was possible to correlate paralysis and curvature. In the cases of thoracic scoliosis there were a few patients with normal intercostals. In the thoraco lumbar curvature it was occasionally difficult to know why curvature developed as almost no paralysis existed. Despite these exceptions in the thoracic curves the principal factor would appear to be a paralysis of intercostal muscles and in the thoraco lumbar curves a combination of weak lower intercostals and lateral abdominal flexors on the convex side. Particularly in the thoraco lumbar curves the effect of gravity in the presence of a symmetrical severe paralysis was easily seen.

It is concluded that the most important cause of paralytic scoliosis is asymmetrical paralysis of lateral trunk flexor muscles whether intercostal or abdominal. If the consequent asymmetrical muscle pull had many years in which to act upon the growing vertebrae then a severe curve might be expected. If the paralysis was contracted later in childhood only a small curve would appear.

NEUROFIBROMATOSIS

In neurofibromatosis the typical curvature is thoracic and the curve is short usually involving 5 or 6 vertebrae (Fig 78). The curvature is sharp and almost angular and there is often an associated kyphosis. However although the thoracic variety is common there are numerous variants sometimes a lumbar curve or an atypical double primary curve in the thoracic region. The general prognosis of neurofibromatous scoliosis is poor. The development of a severe curve with kyphosis is common and this curve is so sharp and rigid that correction is difficult. Correction and fusion is therefore usually indicated at an early stage.

NEUROPATHIC AND MYOPATHIC SCOLIOSIS

A variety of scoliotic patterns is found in the neurological conditions complicated by scoliosis for instance syringomyelia Charcot Marie Tooth neuropathy and Friedreich's ataxia. In syringomyelia the curve is in the thoracic region. The most characteristic are double primary curves which are unequal and give the child the aspect of decompensation. The aetiology may sometimes be suggested by this imbalanced appearance which is uncommon in the scolioses described previously or suspicion may be aroused when a curve thought to be idiopathic progresses differently from or more rapidly than would be expected. Re examination may disclose early neurological changes.

Occasionally the problem arises whether the scoliosis is primary with the neurological condition secondary to the scoliosis or whether the scoliosis is primarily due to a neurological disease. In the neurological diseases that may simulate paraplegia the differential diagnosis can be difficult and may require myelography to exclude local compression of the spinal cord.

The myopathic scolioses are not of great importance. They may be seen in muscular dystrophy but do not progress notably. They are also a rare complication of arthrogryphosis multiplex congenita.



(a)



(b)

FIG 78 — Neurofibromatosis
 (a) and (b) Several small café
 au lait patches are obvious and
 it is common to find no more
 cutaneous evidence of neuro
 fibromatosis than in this boy
 (c) The short thoracic curve
 of five vertebrae is also
 characteristic



(c)

STRUCTURAL SCOLIOSIS

THORACOGENIC SCOLIOSIS

Thoracogenic scoliosis is now quite uncommon. It was described in relation to chronic empyema in which fibrosis induced a structural curve concave to the fibrotic side.

Scoliosis may be seen after pneumonectomy or thoracoplasty; this also is extremely uncommon. On occasions it is related to the removal not only of the ribs but of the transverse processes (Wenger and Hermann 1941).

KYPHOSCOLIOSIS

In many severe scolioses there appears to be a kyphosis and the term kyphoscoliosis has been much used. At operation it is possible to see the back of the primary curve and in more than 200 such operations it has become obvious that an antero-posterior deformity in association with scoliosis is not common—much less so than would be expected from the clinical appearance.

A true kyphosis may be seen as a part of the curves of congenital and infantile idiopathic kyphoscoliosis and neurofibromatosis.

Congenital kyphoscoliosis

The most important of the kyphoscolioses are undoubtedly congenital, commonly cervico-thoracic or thoraco-lumbar. In 21 cases of congenital kyphoscoliosis the cervico-thoracic lesion was found on 5 occasions and the thoraco-lumbar on 16. In 16 patients confirmation of the congenital aetiology was obtained either by radiography or at operation. In 5 patients early radiographs were wanting and a congenital kyphosis was not finally confirmed but the deformity resembled that in the other 16.

In the early years there is a pure scoliosis; it is only as growth progresses that kyphosis appears, often at 10–15 years of age. Suspicion that a scoliosis is becoming a kyphoscoliosis should be aroused clinically when the posterior projection becomes very obvious; radiologically there is an angulation or 'squaring' of the curvature.

In the upper thoracic region the kyphosis is not obvious; what looks no more than a dowager's hump may be betrayed by the radiograph as a true kyphosis of 90 degrees (Fig. 79).

Thoraco-lumbar kyphoscoliosis is rarely severe in early childhood but may rapidly progress so that a kyphosis of 90 degrees is common and it may approach 150 degrees at the end of growth (Fig. 80). Correction is difficult or impossible.

Paraplegia in children with scoliosis may develop in late childhood or at the end of growth. The scoliosis is almost always congenital and usually a kyphoscoliosis. In kyphoscoliosis the compressing agent is anterior to the spinal cord, a ridge of the apical bodies. Alternatively there may be a congenital band (Dewar 1949), a dermoid or other developmental anomaly.

Idiopathic kyphoscoliosis

Although kyphoscoliosis has been described in idiopathic curvature it has occurred in this series only in infantile idiopathic thoracic scoliosis. In 10 per cent of these curves, if severe, there has been a true kyphotic deformity. This kyphosis

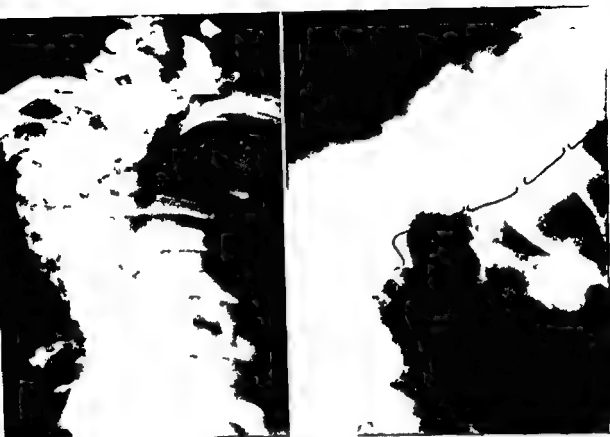


FIG. 79—Cervico thoracic congenital kyphoscoliosis. Congenital lesions could be seen in the early radiographs and were confirmed at the operation done to relieve the paraplegia (B) courtesy of the Editor of *J Bone Jt Surg*

may appear within 2-3 years of the onset of scoliosis and may reach 100 degrees and be as severe a deformity as is the scoliosis. Paraplegia has not been seen in idiopathic kyphoscoliosis.

Neurofibromatosis

Neurofibromatosis as already stated may show a kyphotic deformity as well as a scoliosis. The diagnosis is easily established by the presence of cutaneous pigmentation.

TREATMENT

CONSERVATIVE TREATMENT

It is not proposed to discuss at length the treatment of scoliosis by conservative methods. Although such treatment in its numerous modifications has been used for many years there has as yet been no demonstration of its value. We have abandoned conservative treatment by exercises and orthodox supports because there seems to be sound evidence that it is valueless. It is necessarily time consuming and expensive.

Conservative treatment must, however be considered in children who are too young for operation but have a rapidly increasing curvature—commonly an infantile idiopathic thoracic scoliosis, paralytic scoliosis or rarely a congenital curve.

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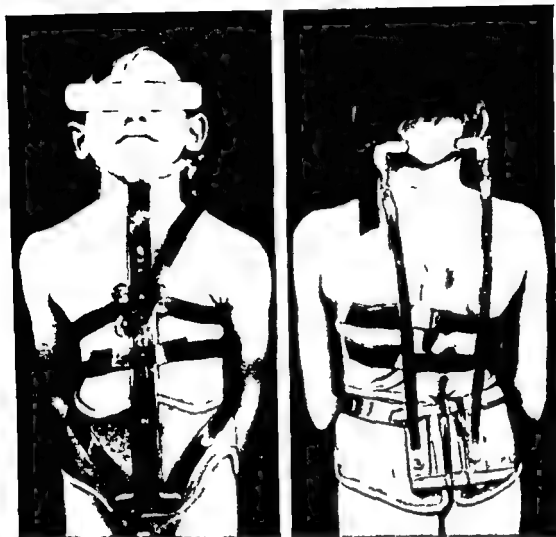


FIG 81 —Milwaukee jacket devised by Walter Blount

surgeons the foremost of whom is Risser perform correction and fusion in very young children. They believe that this is a safe and satisfactory procedure that allows correction of early curves and also prevents progress of the deformity. Others prefer to delay until growth has stopped after which correction and fusion of the mature curve are carried out. Although the maintenance of correction of deformities in growing children is notoriously difficult the correction of mature scoliosis is unsatisfactory. It is unsatisfactory because the curves are by then often gross and rigid and the major deformity of rib rotation can not be corrected. If surgical correction of mature curves is to be the sole method of attack, then the results will always be disappointing.

It seems reasonable to carry out correction and fusion at an age when curvature has not become gross and is still mobile and when rib rotation although moderate is not bad. By delaying correction and fusion until the later years of growth possible recurrence of deformity during many years of growth is avoided. It has become the practice in the Royal National Orthopaedic Hospital to perform correction and fusion usually between the ages of 10 and 12 years.



FIG 80—Congenital thoraco lumbar kyphoscoliosis. There were defects at the eleventh and twelfth thoracic vertebrae and a kyphosis of 125 degrees at the end of growth. No paraplegia. (By courtesy of the Editor of *J Bone Jt Surg*)

The march of events may be so rapid that operation may seem desirable at an early age. Whilst this may be correct it has been our practice to try to delay an increase in deformity until the child is at least 10 years of age. The Milwaukee jacket (Fig 81) which has antigravity distracting turnbuckles and is thus unlike previous supports has proved of value in such cases.

About 50 children from 4 to 8 years of age with progressing curves have been fitted with Milwaukee jackets. Although there has elapsed insufficient time for a definitive report on this problem a firm impression has been gained. In no instance has a curve increased after the Milwaukee jacket has been applied. A few children have obtained and held 40 degrees of correction for as long as 3 years; more often 10–20 degrees of correction has been held, in some little or no correction has been possible but deterioration has been avoided.

It is an uncomfortable jacket and in a minority of cases it produces mandibular recession and protrusion of the upper frontal teeth. It seems therefore essential to use it only for a few years to delay operation.

SURGICAL CORRECTION AND FUSION

Surgical correction and fusion of spinal curvature has been employed for a number of years and the principles and indications are becoming clarified. The age at which correction and fusion is best is still controversial. A number of

Congenital scoliosis

The prognosis of individual curves is difficult and close observation of those cases that are progressive allows intervention at a time before the deformity is gross and when the curve is still mobile. Congenital curves often contain many fused vertebral elements and quickly become rigid and impossible to correct.

Paralytic scoliosis

In paralytic scoliosis the problem is somewhat different. In all patterns the prognosis is worse than in idiopathic scoliosis and operation is carried out for most scolioses that are rapidly progressive in the years of growth. A paralytic curve that measures 60 or 70 degrees at the age of 10 is likely to progress to serious deformity and requires correction. The high thoracic curve requires early correction and fusion because the rib rotation at its upper end cannot be altered by spine fusion.

A need for surgery in paralytic scoliosis may arise from spinal fatigue, back ache and instability, symptoms often suffered by a patient with poor trunk muscles. In the collapsing unstable spine due to symmetrical severe paralysis of the trunk muscles, stabilization of the spine from the mid thoracic region to the sacrum is often valuable. This may change a patient able to walk with difficulty into someone able to walk without a spinal support and thus with improved mobility. Other such patients often have paralysed legs and are unable to walk or even sit up without the support of their arms or a heavy spinal brace. A long spinal fusion for stabilization may convert a poor sitter into a good sitter, able to earn an independent living at a sitting or bench occupation.

In stabilizing a spine by fusing the vertebral column to the pelvis, one important factor must be considered. When a paralysed patient walks, he must lift his legs by hip flexors or by elevating his pelvis with his lateral abdominal flexors. After fusing spine to pelvis it is impossible to use the lateral abdominal flexors and it is possible for a moderate walker to be immobilized in consequence of surgery. Before fusing to the sacrum, therefore, it is essential to determine whether there are hip flexors, and if their power is dubious, a useful test is to apply a closely-fitting plaster spinal jacket. If the patient can walk, fusion to the sacrum is permissible.

Neurofibromatosis

In neurofibromatosis the general prognosis is so poor that correction and fusion are usually indicated. Close observation may indicate its early necessity or the possibility of delay.

In myopathic and neuropathic scoliosis, correction and fusion are rarely indicated because the primary disease is usually more important than the scoliosis and is itself a bar to major surgery.

Congenital kyphoscoliosis

Kyphoscoliosis of the upper or lower thoracic vertebrae may increase rapidly during the later years of childhood and produce paraplegia. As this deformity is a scoliosis and kyphosis, correction is difficult, particularly as there may be congenital vertebral fusions and the curve may be rigid. Correction by distraction jacket may not reduce the deformity enough to relieve the paraplegia.

Indications

When a scoliosis is mature the existing deformity must be dealt with, but the surgeon knows there will be no further change and the existing visible deformity is the only one that concerns him. A mature scoliosis is indicated by the ingrowth of the iliac apophyses.

When considering correction and fusion of a mature curve it is necessary to know what surgery can achieve. By correction we can elevate a dropped shoulder, and a hip can be made less obvious but the deformity of prominent ribs cannot be overcome. If the shoulder and the hip are the obvious deformities, and if the curve is mobile, correction and fusion are indicated. If the curve is rigid or if the shoulder and hip are not prominent correction and fusion will not improve the appearance of the patient, however severe the curve.

In the immature scoliosis there is a more complex problem but one in which a solution is often easier. If a young growing child has a curve that from our study of the curve patterns can be expected to increase to gross deformity then preventive correction and fusion is invaluable. It is believed that the most worthwhile efforts of surgery in the treatment of scoliosis must be based on timely and early intervention in deformities that will increase seriously.

The onset of menstruation and the skeletal age may be used as pointers to the approach of maturation.

Surgery is necessary in only 5-10 per cent of idiopathic curves and approximately 30 per cent of paralytic curves and is discussed below in relation to the various types of scoliosis.

Idiopathic scoliosis

The infantile idiopathic curve almost invariably needs correction and fusion and this is undertaken at the age of 10 years. Often the child has been in a Milwaukee jacket to prevent increasing deformity. Juvenile and adolescent thoracic scolioses often but by no means always require correction and fusion. Thoracic curves of 60-70 degrees at the age of 10 years may be expected to increase so much that correction is needed. In children in whom scoliosis develops later than this close observation will demonstrate which curves are the more progressive and of the adolescent thoracic curves some 40-50 per cent need operation.

Thoraco lumbar curves rarely develop a deformity needing correction and fusion but there are a few in which one hip becomes very prominent even when the curvature is small and these can be recognized by serial observations. Lumbar curves almost never require correction and fusion for cosmetic reasons.

The double primary curves show a very small number of severe deformities. The hip and shoulders do not alter and the main deformities are the double rotation and the shortness of the trunk. In some 5 per cent of these primary curves of more than 100 degrees develop these if discovered early may be corrected and fused. The appearance is not improved and the only reason for operation is the prevention of excessive shortening of the trunk. Fusion of two primary curves is an extensive operation and since it is not finally established that spinal growth continues after fusion the value of intervention must await further study.

CHAPTER 4

DEGENERATIVE DISEASES

DOUGLAS H. COLLINS

INTRODUCTION

THE DEGENERATIVE diseases with which we are here concerned are those that are prominently linked with advancing years and that having no clearly apparent extrinsic causation such as acute injury or infection seem to depend largely upon involutionary or senile changes. They are characterized in the main by destructive changes that are not balanced by the regenerative effects normally invoked in younger tissues and they lead to functional incompetence such as may be exemplified by the loss of the normal texture of the skin in senile elastosis, the rigidity of the wall of a sclerotic artery or the failure of an ageing spinal column to maintain the fully erect posture.

Much has been learnt in recent years about the nature and distinctive types of degenerative diseases of the skeleton mainly through the critical use of standard methods of pathology and radiology. If we cannot yet define the causes of these diseases—and the problem of their causes is no less than a part of the vast biological problem of senescence and mortality of all living matter—we do now know some thing of their circumstances, of how they progress and to what they may lead. In the case of the locomotor system the effects of ageing cannot be dissociated from wear and tear, the results of long exposure to repeated minor trauma. The purpose of this chapter is to describe and illustrate the pathology of degenerative conditions of the spine.

Degenerative processes may affect primarily or mainly (1) the synovial spinal joints, (2) the intervertebral discs, and (3) the bone of the vertebrae. This provides a useful basis for cataloguing the different lesions, although such is the functional unity of the spinal column that the degeneration of one element usually leads to secondary changes in the others.

The degenerative diseases of the vertebral column are classified according to the structure mainly or primarily affected.

- (1) *The synovial joints*: osteoarthritis
- (2) *The intervertebral discs*: degenerations, herniations and protrusions, osteophytosis (spondylosis), senile kyphosis, and adolescent kyphosis
- (3) *The bones*: osteoporosis and Paget's disease

SYNOVIAL JOINTS OF THE SPINE

Osteoarthritis

Osteoarthritis is a disease of the synovial (diarthrodial) joints. Its pathological features are well defined. It begins with a lesion of the hyaline articular cartilage which results in most instances from wear and tear of ageing cartilage. Flaking off of the surface of the cartilage is followed by fibrillation of its depths. Subsequently a series of rather elaborate changes takes place in the joint in the

Congenital scoliosis of the upper or lower thoracic vertebrae should be watched for the appearance of a kyphosis this may indicate an impending paraplegia. Correction followed by fusion is advisable to avoid an ugly deformity sufficient to produce paraplegia. As one of the theoretical disadvantages of early fusion is the development of a lordosis there seems to be little reason for delay in these cases.

If paraplegia persists despite correction cord decompression may be achieved by incision of the dura after laminectomy (McKenzie and Dewar, 1949). This has sometimes led to an increase of the kyphosis from instability and to recurrence of the paraplegia. Incision of the dura should therefore probably be followed by an anterior body fusion. An alternative technique is to relieve the pressure on the spinal cord by an antero lateral decompression. This is a logical operation but the cord is in danger from thrombosis. The correct surgical approach to this problem has yet to be ascertained.

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INTERVETRAL DISCS

In horizontal section a disc is about the same size and shape as the vertebral bodies it unites. Its central part is a soft semifluid mass the nucleus pulposus, which contains over 80 per cent water by weight. The water is bound to the mucopolysaccharides as in myxoid tissues. Whether or not the nucleus is a vestige of the notochord, as was generally thought the disc as a whole appears to have a mesenchymal structure and a mesenchymal function throughout life. Because it contains so much water the nucleus is virtually incompressible and inelastic although subjected to a great range of changing pressures. Surrounding the nucleus are lamellae of fibrocartilage and fibrous tissue the annulus fibrosus containing more cells in abundance of collagen and a lesser amount of mucopoly saccharide ground substance. The annulus is elastic. It blends with the overlying spinal ligaments and its fibres run obliquely from vertebra to vertebra being continuous with the fibrils in the cartilaginous end plates and ossified epiphyseal rings of the vertebral bodies. Annulus and nucleus are most sharply demarcated in childhood and as age advances the distinction between the two is gradually lost (Fig 82).

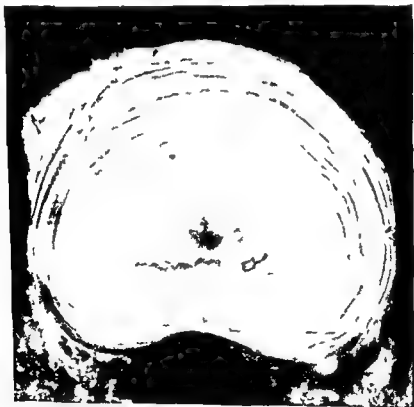


FIG 82—Cross section view of a lower thoracic intervertebral disc of an elderly man with osteophytosis. Note the concentric laminae of the annulus fibrosus. A distinct nucleus pulposus is no longer evident. The centre of the disc is degenerated and fissured. There were large antero lateral osteophytes between which the broken outer fibres of the disc protruded (top left) ($\times 1.5$).

Dehydration with loss of definition of the nucleus pulposus is the principal feature of the degenerations that affect the discs. The disc is avascular. The amount of water that it contains depends on the nature and perhaps on the degree of polymerization of its mucopolysaccharides which have varying hydrophilic

course of which the bone ends become altered in shape and form denuded of cartilage and surrounded by spurs or shelves of new osteophytic bone

This type of disease is well known as an affection of the hip or knee, but it may also affect any synovial articulation large or small and especially those that are subject to free movement under much mechanical strain

The small diarthroses of the vertebral column the posterior apophyseal joints and the central atlanto axial joint, are not exempt The more advanced stages of the disease in the posterior spinal joints have been detected both by radiography and by the examination of macerated anatomical specimens (Shore, 1934 35) and I have elsewhere (Collins 1949) described and illustrated the earlier cartilaginous lesions in these joints

The clinical correlations of this condition are not yet accurately determined because the full range of movement is less easily tested in these joints than in the larger limb joints and radiographic subtleties like 'loss of joint space' that is thinning of cartilage can hardly be shown Nevertheless it is important to know of this condition It is quite distinct from osteophytosis of the spine the more familiar lesion of bony spurs arising from the vertebral bodies that is dependent as we shall see on changes in the intervertebral discs

Some authors (for example Cave Griffiths and Whiteley 1955) have described, as a cause of cervical spinal nerve root irritation an osteoarthritis of the neuro central joints of Luschka * But whether these are diarthrodial joints and therefore subject to true osteoarthritis is debatable The marginal osteophytes that form round these 'half joints' and may encroach upon the intervertebral foramen are much like those of the more general spinal osteophytosis and it is not certain that this condition should be separated from the type of spondylosis that depends ultimately on disc degeneration Morton (1950), in a careful radiographic and dissection study of the cervical spine does not specially mention the Luschka joints but he demonstrates how enlargement at an arthritic apophyseal joint may combine with osteophytes arising from the postero lateral lips of the vertebral body to cause compression of a cervical nerve root as it passes through an intervertebral foramen

INTERVERTEBRAL DISCS

Degenerations

The intervertebral discs unite the bodies of successive vertebrae from the second cervical down to the sacrum They form a series of amphiarthrodial or slightly movable joints that possess no synovial cavity or synovial membrane, so that the diseases to which they are subject are fundamentally different from those that affect the synovial joints

About a quarter of the length of the vertebral column of a young adult is made up of the discs The loss of stature in old age is caused partly by thinning of the discs and partly by the usual failure to maintain the fully erect posture To some degree these two factors are interdependent because degeneration of the discs with loss of bulk inevitably tends to kyphosis

* The postero lateral portion of each vertebral body is ossified from the centre for the vertebral arch (the neurocentrum) In the cervical spine below the axis the part of the body usually articulates with the infero lateral aspect of the vertebral body above forming the joint of Luschka which thus constitutes part of the antero medial wall of the cervical intervertebral foramina

spontaneously. So called spontaneous ruptures may, of course, take place at moments of particular mechanical stress but in many instances there is no history of any such precipitating incident.

Direct injury

The direct injury that leads to an escape of the nucleus may be a fracture dislocation of the spine. Sometimes it may be the accidental needling of a disc during the operation of lumbar puncture. This is a real risk (Dripps and Vandam, 1952) as such well documented cases as those of Milward and Groot (1936) have shown. Although serious damage may be rare, especially if the needle is fine. The deliberate acupuncture and injection of opaque fluid into a disc for radiographic purposes (Erlacher, 1952) seems, however, to be a completely unjustifiable procedure.

Spontaneous herniation

Spontaneous herniation of the intervertebral disc may take place either vertically or horizontally and involves an escape of material from the semisolid nucleus pulposus into the spongiosa of one or both of the adjacent vertebral bodies or its escape through the annulus fibrosus, usually backwards into the epidural tissues. In either case prolapse of nuclear substance can take place only when the nucleus retains its turgescence. It cannot occur in old people whose nuclei pulposi have undergone desiccation. This type of acute nuclear herniation is therefore prone to occur in relatively young people even in adolescence. It may be precipitated by injury or strain but there is seldom conclusive evidence about this. It may be that a congenital or acquired defect of the cartilaginous end plate or of the annulus is also a necessary factor.

Vertical prolapse—As illustrated in Fig. 84 vertical prolapse gives rise to what is often called the Schmorl's node—that is a tongue or club shaped mass of white disc tissue projecting into the red spongy substance of the vertebral body. It is seldom more than 1 centimetre across and is continuous with the main part of the disc. It usually lies near the central axis of the vertebra and is therefore most often demonstrable in a post mortem specimen that has been sawn vertically down the central axis. A Schmorl's node cannot be detected radiographically until it becomes outlined by a shell of reactive bone that casts itself around the old prolapse (Fig. 85). The Schmorl's node *per se* is of no clinical importance. It may happen explosively, and it is tempting to think that certain hyperacute attacks of lumbago may be so caused but of this we have no proof. The lesion is an exceedingly common one. Schmorl himself discovered nodes in 38 per cent of spines. They are very often multiple. The important consequence of a vertical herniation of the nucleus is the loss of disc substance and the approximation of the two adjoining vertebral bodies. Multiple Schmorl's nodes are sometimes seen arising from a succession of discs in athletic young persons and are often found in adolescent kyphosis (see page 113). Vertical prolapse involves herniation of nucleus pulposus only, there is no annulus at the central area of the cartilaginous end plate.

properties. Though it is simple to regard loss of water as the most important feature of age changes in the disc, the process is in fact a true degeneration in the biological sense—that is, a change in the composition of a tissue so that it assumes a functionally less efficient form.

Degeneration of disc tissue is accelerated if the disc becomes vascularized, as may happen if the cartilage end plates are damaged. In elderly people fissures appear in the centre of the discs, and an unidentified brownish pigment may be found—the so-called brown degeneration of the discs. Calcification of degenerated discs is sometimes seen and may be detected radiologically (Fig. 83). This is usually amorphous calcification in the central areas of the disc. Actual ossification is seen only as a consequence of organization after fracture of the enclosing bones or cartilage end plates, or near the margins, in the bony bridges of ankylosing spondylitis. Except where calcification has occurred, the only radiographic sign of disc degeneration is the narrowing of the intervertebral space and very often a slight forward tilting together of the adjoining vertebrae.



FIG. 83—Calcification of the intervertebral disc: lateral radiograph of the lumbar spine of an elderly man. Calcification is well seen in the disc L2/3 in centre of picture. There is calcified atheroma of the aorta anteriorly. (B, courtesy of Dr J. Wilkie, Department of Radiology, Royal Infirmary, Sheffield.)

Herniations and protrusions

The nucleus pulposus of an intervertebral disc is constantly under compression. Although not elastic, its turgescence can be demonstrated even after death by the way it bulges when the spine of a young person is sawn open. The turgid nucleus in the living is bounded laterally by the strong elastic lamellae of the annulus fibrosus and vertically by the cartilaginous end plate of the disc. This is a plate of well-developed hyaline cartilage countersunk slightly into the vertebral body and ringed by a rim of bone that has developed from the ring epiphysis of the vertebra.

Escape of the nucleus pulposus may occur either as a result of direct injury or

spontaneously. So called spontaneous ruptures may, of course, take place at moments of particular mechanical stress but in many instances there is no history of any such precipitating incident.

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FIG 84 —Section of a recent vertical prolapse (herniation) of nucleus pulposus of a disc into the spongiosa of a vertebral body. This is the first stage of a Schmorl's node (Haematoxylin and eosin $\times 45$).



FIG 85 —Section showing a later stage of a Schmorl's node. The tongue of disc projecting into the spongiosa of the vertebra has become invested by a thin shell of bone. It is this bony reaction that casts a shadow in the radiograph (Haematoxylin and eosin $\times 25$).

Horizontal prolapse—The disc may suffer two forms of horizontal prolapse (Brain 1954 Frykholm 1951), namely nuclear herniations and annular protrusions. Nuclear herniation the ejection of the soft nucleus pulposus generally takes place only posteriorly but annular prolapse the convex bulging of relaxed annulus fibres occurs in almost any horizontal direction. The author has confirmed these views by the histological examination of much material surgically removed after laminectomy. From the more acute lesions the prolapsed disc tissue was sometimes fibrocartilage of the annulus and sometimes soft myxoid tissue with stellate or balloon cells characteristic of the nucleus pulposus. In the case of more chronic prolapses secondary degenerative changes calcification or invasion by vascular granulation tissue sometimes with later ossification masked the original nature of the herniation.

Posterior herniation of nuclear substance is seldom accompanied by any gross breach of the annulus the semifluid matter seems to escape by dissecting its way through the annulus fibres. Anatomical inspection of vertebral bodies and discs shows the posterior longitudinal ligament to have a scalloped outline its broadest expansions lying over each disc to which its fibres are firmly attached. Most posterior herniations of the disc lie, therefore to one side of the midline and often a little above or below the central horizontal plane of the disc. Neurological symptoms caused by direct pressure may thus be unilateral. The protrusion usually takes the form of a small sessile or nipple like tumour covered by areolar tissue and pressing against the dura mater to which it does not adhere until organization and reactive fibrosis have occurred. Necropsy examination in adults often reveals posterior prolapses of some form over the length of the spinal column although symptoms seem to arise only in the cervical and lumbar regions.

Annular protrusions follow the narrowing of the disc and the telescoping of the intervertebral space that result from the desiccation or vertical prolapse of the nucleus pulposus. It is the relaxed annulus fibres that are as it were squeezed out. Now such backward protrusions may cause symptoms of the disc syndrome but they appear much more commonly between the antero lateral lateral and postero lateral borders of the vertebral bodies. In the postero lateral position they or the bony osteophytes that grow to enclose them may partially obstruct the intervertebral foramina and lead to the radicular symptoms of spondylosis. However the largest annular protrusions are seen anteriorly not in the midline because the anterior common spinal ligament is very strong and very tightly bound to both bones and discs. It is these antero lateral protrusions of annulus fibrosus that account for the common and well known forms of marginalipping of the vertebral bodies or osteophytosis of the spine.

Osteophytosis (spondylosis deformans)

An osteophyte means a bony outgrowth or excrescence. The term particularly designates the small and irregular spurs or shelves that appear at the margins of those limb joints that in later life have become affected by osteoarthritis. In this situation osteophytes are an integral part of the osteoarthritic process in diarthrodial synovial joints. In other situations however osteophytes may be quite independent of osteoarthritis. The vertebral column provides a good

example of this Osteoarthritis may affect the apophyseal joints which are synovial and truly osteoarthritic osteophytes may form around the small articulations but osteophytes that form and sometimes grow to a great size (Fig 86) at the margins of the vertebral bodies are not osteoarthritic although they are often so called. They are the consequence of collapse of the intervertebral discs and protrusion of the annulus



FIG 86 — Osteophytosis (spondylosis deformans) of the lumbar vertebral bodies. Antero posterior radiograph showing very large antero lateral osteophytes. Some appear to have fused but in others the clear area of interposed herniated disc tissue is still apparent (B) courtesy of Dr J Wilkie, Department of Radiology, Royal Infirmary, Sheffield).

Pathogenesis

The mechanism will be explained by reference to Fig 87

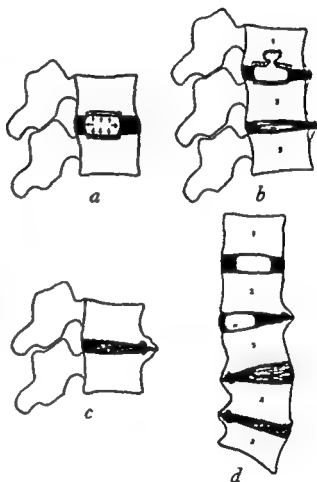
Two vertebrae separated by a healthy intervertebral disc are depicted in Fig 87a. The turgid nucleus pulposus (represented by the clear area in the centre of the disc) normally maintains the separation of the vertebral bodies and gives resilience to the whole disc by virtue of its resistance to compression (arrows). The nucleus pulposus is bounded by the fibrocartilage and collagenous fibrous tissue of the annulus fibrosus (black area) and by the end plates of hyaline cartilage (cross hatched areas) which are countersunk into the vertebrae and surrounded by a ring of bone which was originally the epiphyseal ring and in which the fibres of the annulus are anchored.

Three vertebrae with different conditions affecting the two discs and causing their collapse are shown in Fig 87b. The disc between vertebrae 1 and 2 is narrowed by the loss of a proportion of the nucleus pulposus into the spongiosa of the first vertebral body. This has burst through a defect of the upper cartilage end plate and the

INTERVERTEBRAL DISCS

extruded nuclear substance being outlined by reactive bone would appear both radiologically and in anatomical section as a typical Schmorl's node. The disc between 2 and 3 has collapsed through degeneration. The hyaline cartilage of the end plates has also degenerated. The disc has undergone dehydration and loss of bulk and the original position of the nucleus is represented only by cracks and fissures. In each instance narrowing of the intervertebral space has resulted in slight forward tilting of the vertebrae around the fixed axis of the apophyseal joints. This oblique compression of the discs causes a slight forward protrusion of whatever plastic disc substance still remains comprising in most instances the relaxed anterior fibres of the annulus.

FIG. 87 — Diagrams illustrating stages in the pathogenesis of spinal osteophytes (see text). (a) Normal intact disc. (b) Disc 1-2 is narrowed by a vertical prolapse and disc 2-3 is narrowed by degeneration. In each case the vertebral bodies are tilted slightly forwards and the relaxed annulus fibres bulge forwards raising the overlying periosteal membrane. (c) Ossification within the area of elevated periosteum on either side of the protruded annulus has formed characteristic osteophytes. (d) The lateral tilting of the vertebral bodies in scoliosis leads in the same way to osteophytes on the concave parts of the spinal curvature. This diagram shows an anterior view of the spine. Diagrams a-c show a lateral view.



As the anterior margin of the disc slowly protrudes it carries with it a tent like elevation of the periosteum from the surface of the adjoining vertebral bodies. It is in these areas of periosteal elevation that new bone forms and it is thus that spinal osteophytes develop.

The final development of characteristic kissing osteophytes around a degenerated collapsed and extruded disc is shown by Fig. 87c. It should be compared with Fig. 88 which is a photomicrograph of a section through two vertebrae bearing huge osteophytes of this type. It is important to remember that the clear area seen in the radiograph between two adjoining osteophytes is always occupied by disc substance. It is a matter of common observation that spinal osteophytes hardly ever appear in the midline anteriorly but do so commonly in the antero-lateral position just to the side of the anterior common ligament. The attachment of the anterior common ligament to the

bone of the vertebral bodies is probably so firm that it cannot be displaced. Periosteal elevation and subsequent subperiosteal bone growth on the anterior rim of the vertebral bodies occur therefore to one side or both sides of this ligament.

In Fig 87*d* the truth of this explanation of the origin of spinal osteophytes is also convincingly demonstrated in the scoliotic spine. Osteophytes develop only in the concavities where they are invariably present in every scoliosis that has been fixed for any length of time. The diagram illustrates their development. The disc between 1 and 2 is normal. The disc between 2 and 3 is distorted by the tilting of the vertebral bodies. Such tilting in the early stages causes a displacement of the nucleus pulposus towards the wider part of the disc on the convex surface of the curvature with the extrusion of disc tissue in the concavity and the growth of osteophytes around it. The discs between 3 and 4 and between 4 and 5 show the degeneration or fissuring usually seen in sawing through such a spine and the development of osteophytes again within the concavity. In long standing scoliosis there is almost always a secondary distortion of the vertebral bodies which become either rhomboid or wedge shaped (vertebrae 4 and 5). Alterations in the form of both discs and vertebrae perpetuate the deformity of the spine.

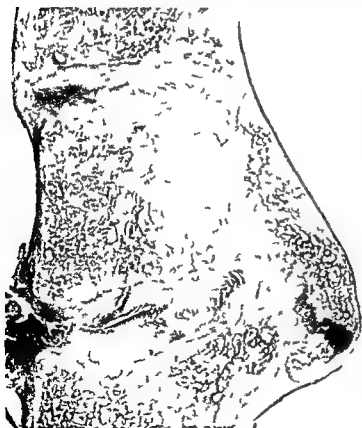


FIG 88—Large section in paramedial sagittal plane of three cervical vertebrae. The upper disc is healthy. The lower is degenerated and protruded between the two very large kissing osteophytes. These are formed of well developed bone applied as can be seen in the case of the lower one to the outside of the original wall of the vertebral body (Celloidin section. Haematoxylin and eosin $\times 28$).

The two halves of the thoraco lumbar portion of the spine of an elderly man with much osteophytosis are shown in Figs 89 and 90. The left half (Fig 89) has been macerated and the right half (Fig 90) retains the soft tissues including discs and ligaments. These preparations clearly demonstrate the true nature of vertebral body osteophytes and their dependence upon vertical collapse and horizontal bulging of the discs. This specimen showed no evidence of osteoarthritis.

INTERVERTEBRAL DISCS

of the synovial joints of the spine thus emphasizing the distinction between osteoarthritis and osteophytosis

Only the antero lateral osteophytes have so far been discussed. These are the common ones because almost all the forces acting upon the adult spine tend to promote a forward bending but occasionally the annulus protrudes backwards towards the spinal canal though seldom very far, and small osteophytes form around such protrusions in exactly the same way. Posterior osteophytes may, of course, develop on either side of a long standing posterior prolapse of the



FIG 89—The left half of a spine with severe osteophytosis. Vertebrae T9-L2. After maceration the separated dry bones were reset in their correct anatomical position. Note the large anterior osteophytes and the slight kyphosis.



FIG 90—The right half of the same specimen as shown in Fig 89. Note the degeneration and narrowing of each of the discs associated with osteophytes and its forward protrusion between the new bony spurs.

nucleus pulposus but most clinical importance attaches to those that form in the postero lateral position and press upon the spinal roots in their course either through the intervertebral foramina or across the spinal canal. These may account for certain of the neurological symptoms associated with spondylosis.

Spondylosis

There is some confusion about the term spondylosis which should be resolved. The suffix *sis* or *osis* should never have been appended to a normal anatomical term like *spondylo* which simply means *vertebra*. Spondylosis by

bone of the vertebral bodies is probably so firm that it cannot be displaced. Periosteal elevation and subsequent subperiosteal bone growth on the anterior rim of the vertebral bodies occur therefore to one side or both sides of this ligament.

In Fig 87d the truth of this explanation of the origin of spinal osteophytes is also convincingly demonstrated in the scoliotic spine. Osteophytes develop only in the concavities where they are invariably present in every scoliosis that has been fixed for any length of time. The diagram illustrates their development. The disc between 1 and 2 is normal. The disc between 2 and 3 is distorted by the tilting of the vertebral bodies. Such tilting in the early stages causes a displacement of the nucleus pulposus towards the wider part of the disc on the convex surface of the curvature with the extrusion of disc tissue in the concavity and the growth of osteophytes around it. The discs between 3 and 4 and between 4 and 5 show the degeneration or fissuring usually seen in sawing through such a spine and the development of osteophytes again within the concavity. In long standing scoliosis there is almost always a secondary distortion of the vertebral bodies which become either rhomboid or wedge shaped (vertebrae 4 and 5). Alterations in the form of both discs and vertebrae perpetuate the deformity of the spine.



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end plates and are quite distinct from the ligamentous and marginal body bridges of ankylosing spondylitis. In addition to degeneration vertical or antero-lateral prolapse of the discs may contribute to their narrowing and Schmorl's nodes and antero-lateral osteophytes may be seen.

The vertebral bodies are atrophic and may show some osteoporosis but since the discs first lose their turgescence the bi-concave fish-vertebra deformity characteristic of senile osteoporosis is not seen. The vertebral bodies tend however to become compressed in front and their slight wedge shape adds to the general kyphotic deformity.

Adolescent kyphosis

This deformity is an exaggeration of the dorsal curve and extends often through the lower thoracic (dorsal) and lumbar region. It is known sometimes as Scheuermann's disease and because of the often ragged or fragmented appearance on the lateral radiograph of the upper and lower margins of the vertebral body (Fig 91) it is often considered to be an epiphysitis or osteochondritis.



FIG 91—Adolescent kyphosis. Lateral radiograph showing characteristically ragged surfaces of the vertebrae, some Schmorl's nodes and considerable kyphosis. (By courtesy of Dr J Wilkie, Department of Radiology, Royal Infirmary, Sheffield.)

When these spines are examined anatomically an almost continuous series of vertical prolapses of nucleus pulposus (Schmorl's nodes) is seen (Schmorl and Junghans 1953). Nuclear herniations are the primary lesion although abnormality of or injury to the cartilage end plates may predispose to them. The discs collapse and forward tilting of the vertebrae follows. The integrity of the discs is lost early in the disease. The integrity of the bones also may be lost unless preserved by corrective or supportive treatment because pressure interference with their normal growth may lead to severe wedge deformity of the vertebral bodies and consequent increase in the kyphosis.

derivation strictly implies no more than the state of having vertebrae Osteophytosis, on the other hand denotes the state of having osteophytes which is meaningful As a descriptive pathological term therefore, osteophytosis, or spinal osteophytosis, is to be preferred Schmorl however, introduced spondylosis deformans as a better substitute for the older term spondylitis deformans, because he knew that the condition was non inflammatory The term certainly renounces this concept and being less specifically descriptive than osteophytosis, it may be held to include also the disc degeneration and collapse that underlie the osteophytes Whatever the pathological state is called—the author still prefers osteophytosis— a term is needed to designate that complex clinical syndrome of subjective symptoms and objective neurological signs that is associated with a rather small proportion of lesions and spondylosis seems to be filling this need Let us therefore, agree to call the clinical syndrome spondylosis and the pathological condition spinal osteophytosis We may thus establish an analogy with nephrosis, which has come to have a clinical connotation more distinctive than its pathological

Spondylosis then is the clinical syndrome arising from an acquired deformity of the spine in which the principal pathological lesions are narrowing degeneration or prolapse of one or more intervertebral discs with osteophytosis of the related vertebral bodies The clinical aspects of the matter are discussed elsewhere in this volume (see Chapter 9) but it may be useful here to recall the observation of Inman and Saunders (1947) that the intervertebral disc is primarily a part of the functional mechanism of the spinal column and that its relationship to the spinal cord and the nerve roots is secondary and fortuitous In derangements of the disc these authors go on to say, one is dealing with a dual problem, first deranged spinal mechanics that may give rise to disability and pain and secondly the effects of spinal nerve or cord irritation and compression that may include not only pain but also objective neurological signs Brain (1954) who with Frykholm believes that the nerve roots in cervical spondylosis become involved through a reactive fibrosis of the dural nerve root sleeve, points out that, in addition to direct compression by osteophytes or protruded disc and in addition to the abnormally limited movement of the affected part disc collapse leads to a shortening of the cervical spine thus disturbing the relationship between the radicular nerves and their corresponding foramina

Senile kyphosis

This rather common deformity of the elderly is a smooth kyphotic curvature of the upper and middle parts of the thoracic spine usually with a straightening out of the normal lumbar curve In its advanced form senile kyphosis causes the pronounced round shouldered stoop and forward thrust of the neck that sometimes accompanies the general wasting and loss of stature of the old of either sex The deformity is apparently painless It soon becomes fixed through ossification across the narrowed anterior parts of the discs

Sagittal section of the thoracic spine reveals the changes that affect primarily the discs and secondarily the bone The discs are degenerated severely atrophic and narrowed especially in their anterior parts Brown degeneration is common and bony bridges sometimes traverse the narrowest parts of the disc joining the spongiosa of one vertebral body to the next They are caused by penetration of vascular granulation tissue from the broken down anterior parts of the cartilage



FIG 92—Normal vertebral body after maceration. The normal texture of the spongiosa is one of a system of rather thick trabeculae and small plates of bone (x 45) (Inset radiograph of the specimen)

A good comparison between the normal vertebral spongiosa and that of the porotic vertebra is seen after maceration of the bones, that is after removal of organic soft tissues by caustic solutions or proteolytic enzymes (Figs 92 and 93). Here it is seen that the normal strong system of little bony plates has been reduced to a delicate web of thin struts.

Cooke (1955) in a full discussion of many aspects of osteoporosis described the great strength of the vertebral centra that normally can resist a crushing force of some 600–900 pounds per square inch. The porotic vertebrae he tested were able to withstand some 300 pounds or less per square inch. It is now well known that patients with considerable osteoporosis may suffer fracture of several vertebrae with slight or negligible injury. The author's experience, however in examining many spines in the post mortem room is that old fractures are rarely seen although osteoporosis is quite common.

The well known fish vertebra deformity arising from the biconcave compression of the vertebral bodies by expansion of the intervertebral discs is quite often seen in the necropsy room as in the radiological department (Fig 94) but this is not an inevitable consequence. It cannot occur unless the discs retain their turgescence, and there are many cases in which the vertebrae become porotic after the discs are already degenerated and narrowed.

It is generally accepted that increasing age and sexual involution are important factors in the development of osteoporosis. The disease is commoner in the female sex, occurring four times more often in women than in men in the series of Kesson, Morris and McCutcheon (1947) and six times more often in Cooke's (1955) cases. Albright has called it post menopausal osteoporosis in order to emphasize

THE BONES OF THE SPINAL COLUMN

Vertebral bone is by far the most interesting bony territory in the body, quite apart from its articulations and discs and its functions as the axis of the skeleton and the protector of the spinal cord. It contains most of the haemopoietic marrow in adult life, and its diseases and those of blood formation are often intimately related. The vertebrae are highly vascular, and perhaps on this account they are the commonest skeletal site for the growth of embolic metastatic cancer. Though secondary tumours are common, primary bone sarcomas of the spine are rare. Nevertheless the vertebrae often contain lesions of such diffuse neoplasms as myeloma, Hodgkin's disease and lymphosarcoma.

Another special character of vertebral bone is that it is almost wholly cancellous. Having through its trabecular structure, a huge area of endosteal surface it can display great cellular activity. It can resorb, regenerate and remodel itself quickly and like the cancellous bone in the epiphyses of long bones it has a much more rapid metabolic turnover than compact bone. Degenerative processes such as osteoporosis therefore become manifest first and foremost in the vertebral column. The lesions of Paget's disease are also most often to be found there.

Osteoporosis

The term osteoporosis has a precise meaning in bone pathology. It means a quiet atrophy of bone due to a failure of regeneration rather than to an accelerated rate of destruction (Collins 1949). In osteoporosis there is a reduction of the mineral content of a bone *pari passu* with a diminution in size of the bone mass. The trabeculae of the spongiosa first show atrophy. Changes in cortical or compact bone, the widening of the haversian canals and marrow spaces and the thinning of cortical plates, occur more slowly.

A moderate degree of osteoporosis, not confined to the vertebral column, is as common an accompaniment of increasing age as atrophy of the skin and subcutaneous tissues or of the liver, but in a more severe degree senile osteoporosis notably affects the spongy bones of the spine and may cause clinical symptoms. These and the metabolic associations of osteoporosis are dealt with in Chapter 5. All that is necessary here is to emphasize its pathological features and relationships.

Comparative pathological features

Faced with histological preparations of osteoporotic bone, the pathologist is unable to distinguish between senile osteoporosis and disuse atrophy. What he sees is bone tissue, normally staining but composed of trabeculae and lamellar sheets that are thinned and haversian canals that are widened. There is no endosteal cellular activity. There is neither enhancement of osteoclastic resorption nor evidence of new bone deposition by plump active looking osteoblasts. There are no osteoid borders. What bone there is appears otherwise normal and in undecalcified preparations is seen to be calcified normally. The histological appearances in fact confirm the view that osteoporosis is caused by failure of the osteoblasts to restore bone that has been lost in the normal cycle of resorption. That resorption is not accelerated is borne out by the fact that as in normal bone it is only by chance that the histologist sees any osteoclasts, and there is never any area of osteitis fibrosa. The marrow elements in the porotic bones are quite unaffected.

FIG. 94 — Senile osteoporosis. Lateral radiograph of lumbar spine showing advanced rarefaction and biconcave deformity (fish vertebra) of the vertebral bodies. There is compression of the body of lumbar vertebra 1 and a straightening out of the lumbar curve. (By courtesy of Dr J. Wilkie, Department of Radiology, Royal Infirmary, Sheffield.)



TABLE I
INCIDENCE OF OSTEOPOROSIS OF VERTEBRAL CANCELLOUS BONE

<i>Hospital necropsies on patients aged 40 years or over</i>	<i>Cases examined</i>	<i>(++) S.G. < 1.025</i>	<i>(+) S.G. 1.025-1.050</i>	<i>(+ and ++) S.G. < 1.050</i>
Men	189	2	13	15 = 8 per cent
Women	147	4	22	26 = 18 per cent
Both sexes	336	6	35	41 = 12 per cent

By the same yardstick the incidence of vertebral osteoporosis (both sexes) was found to increase with age as shown in Table II

TABLE II
INCREASE OF VERTEBRAL OSTEOPOROSIS WITH AGE

<i>Age group</i>	<i>Number of cases</i>	<i>Per cent</i>
40-49	0 in 45	0
50-59	1 in 78	1
60-69	7 in 99	7
70-79	20 in 94	21
80-89	12 in 19	63
90-99	1 in 1	100

Histological examination of many specimens showed them all to be examples of simple osteoporosis and the figures indicate its great frequency in the ageing population

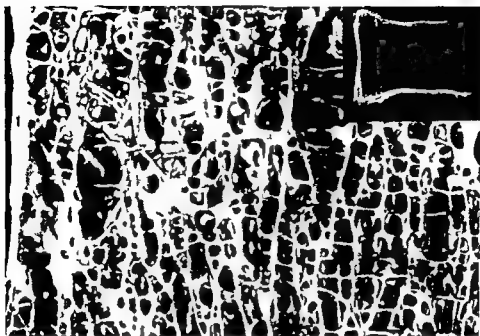


FIG 93—Osteoporotic vertebral body after maceration. On comparison with Fig 91 the reduction of the mass of bone is clearly seen. There is here a more open meshwork of delicate trabeculae enclosing some rather large pools of marrow and there are very few plates of bone ($\times 4.5$) (Inset: radiograph of the specimen).

what he considered to be one of the main factors in its aetiology (Albright, Smith and Richardson, 1941)

Incidence

Some years ago the author made an assessment of the incidence of osteoporosis at necropsy by examining the bone of the lumbar vertebral bodies in an unselected series of necropsies.

The definition of osteoporosis in terms of gross morbid appearances and its recognition in the post mortem room for statistical purposes proved rather difficult. A general description might be that the cancellous bone of the vertebral body was noticeably light in weight, could be minced up easily with scissors and, on scrubbing the surface or after maceration of the bone, the cancellum was seen generally to have an abnormally wide meshwork. Bone like this was markedly rarefied radiographically and the discs showed biconvex expansion unless they were dehydrated or degenerated.

Various attempts to measure the actual density of the bone were made and finally the practice of cutting out small cubes of cancellous bone from vertebral centra and dropping them into copper sulphate solutions covering a range of specific gravities was adopted. The method is a crude one but the results are of some interest. In spite of the variables that attach to the presence of fatty and cellular marrow the main reason for the increased density of bone over other tissues is its mineralization.

Normal bone has a specific gravity (S.G.) of 1.075. The S.G. of some doubtfully pathological bone (\pm) lay between 1.050 and 1.075. Porotic bone (+) had an S.G. of less than 1.050 and a few specimens of very soft bone (++) floated even at 1.025.

The incidence of osteoporosis so measured worked out as shown in Table I.

3 out of every 4 cases of the disease. It is probable that Paget's disease remains throughout life in a subclinical form in about two thirds of the cases. In view of the great frequency of the disease and of its prevalence in subclinical form the incidence of fracture and of sarcoma as complications have always been much over estimated. None the less, when to these we add the much larger number of instances where Paget's disease leads to pain or deformity, it clearly constitutes a medical problem of considerable importance among an ageing community.



FIG. 96.—Isolated lesion of Paget's disease in fourth lumbar vertebra. Radiograph of a necropsy specimen of the half vertebral bodies showing the box-like pattern of sclerosis with a more rarefied centrum.

Paget's disease may involve any part of the bones of the vertebral column—bodies, spines or laminae—but it is in the bodies that the processes of the disease are best seen. Sometimes a succession of vertebral bodies may be affected (Fig. 95) but affection of a single vertebra is not unusual (Fig. 96). This single affection was found by Delitala (1946) in 10 of 110 cases studied radiographically. The author also found solitary foci of the disease in about 10 per cent of cases sometimes in a single vertebra but sometimes in the skull or elsewhere. Such a lesion, the so-called monostotic or mono osteitic form of the disease, does not

Paget's disease

Paget's disease of bone (osteitis deformans), almost certainly should not be included in the category of degenerative diseases but it is conveniently discussed here. It is degenerative only in so far as it is markedly age linked, being common among persons of both sexes past middle age, but the vigour of the cellular reactions that lead to such a change and the enlargement of the bone mass that may result put the disease rather in the category of a hyperplasia.

Paget's disease affects the vertebral column especially in the lumbosacral region, more commonly than any other part of the skeleton. The author found lesions there in 76 per cent of the 46 cases where he made full necropsies.

Lesions are not confined to the lower part of the spine but may occur in any of the vertebral bones from the atlas downwards. In a recently published survey (Collins 1956) the author showed in the detailed examination of the bones in 650 unselected necropsies an incidence of lesions of Paget's disease of 3.7 per cent among patients aged over 40 years dying in hospital. This result compares with Schmorl's (1932) figure of 3 per cent and confirms that a very large number of elderly people have foci of the disease somewhere in the skeleton.

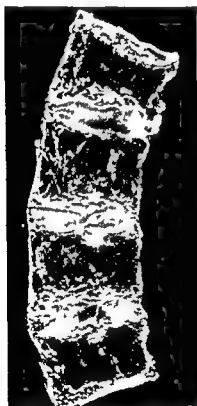


FIG 95—Paget's disease affecting four lumbar vertebral bodies. Osteolytic phase is predominant. The centra are excavated and contain pools of red marrow traversed by a few stout vertical trabeculae. Sclerosis of the walls and end plates is not yet much of a feature. Vertical disc prolapses are seen in connection with the lower two discs.

Further results presented in the same paper were compiled from data derived from a total of 70 cases of Paget's disease pathologically studied. They may be summarized here. About 9 out of every 10 cases are found in persons aged at least 55 years. The sexes are affected probably in the proportion of about 4 men to 3 women. The lumbar vertebrae and sacrum seem to be involved in at least

follows the reconstruction of lamellar bone. But this lamellar bone is subjected to rapidly changing phases of osteoclastic resorption and osteoblastic regeneration and appositional growth. The result is that the bone that is finally elaborated is made up of a sort of mosaic of ill orientated pieces of bone. Each piece is joined to the next by wavy 'cement lines,' narrow bands of metachromatic ground substance that contains no fibres (Figs 99 and 100). Each piece has its own fibrillar or lamellar pattern, and often the fibres may lie in a direction sharply

FIG 98—Paget's disease of lumbar spine and sacrum. Antero posterior radiograph (By courtesy of Dr J Wilkie, Department of Radiology, Royal Infirmary, Sheffield)



at variance with those of the adjacent pieces of bone (Fig 101). These are the features that give what is known as the mosaic pattern to the bone of Paget's disease, and this is the histological hall mark of the disease, stamped on every area of lamellar bone that eventually develops in replacement of the original bony structure.

Clinical features

The classical advanced case of the disease, as illustrated by Paget 80 years ago, with the collapsed and kyphotic vertebral column, enlarged cranium and bowed femora and tibiae, is very rare. Much commoner among the relatively

necessarily mark an early stage in Paget's disease, since the solitary focus may be in a late stage of pathological evolution. A solitary focus simply represents a limited involvement of the skeleton. It is an impressive example of what Albright and Reifenstein (1948) and others have been at pains to point out, that Paget's disease is never a generalized bone disease. Although the lesions may be multiple or even in rare cases quite widely disseminated, they remain focal, and as a rule, the greater part of the bone throughout the skeleton shows no pathological change whatsoever.



FIG. 97.—Paget's disease. Vertebral bone after maceration. The coarseness of the tracery of trabeculae in the spongy bone that develops in this disease is emphasized by comparison with Figs 92 and 93 ($\times 4.5$) (Inset radio-graph of the specimen.)

The coarse tracery of massive trabeculae that develops in the bone affected by Paget's disease is well shown in Fig. 97 which may be compared with Figs 92 and 93. It is this rough scaffolding that gives rise to the well known radiographic appearance (Fig. 98).

Microscopical changes

The microscopical changes that occur in the bone in Paget's disease begin with the osteoclastic resorption of existing bone. This is quickly followed by the active proliferation of primitive coarse fibred bone in a bed of cellular fibrous tissue that replaces previous adipose or haemopoietic marrow tissue. In its wake there

Radiological changes

The radiological diagnosis of Paget's disease in the spine may be uncertain. The radiographs during certain phases of the disease may show hazy or blotchy shadows representing jumbled areas of osteolysis and regeneration. It is only in the later stages of evolution of the disease in a bone that the characteristic coarsely striated x-ray pattern appears, often to be followed by an increasing degree of sclerosis. In the earlier phases the diagnosis may rest between Paget's disease or osseous metastases of carcinoma. Some help may be got from the radiological examination of other areas of the skeleton. The ribs are not very often affected.



FIG. 101. —Section of bone in Paget's disease viewed in polarized light to show the various axes of fibre orientation in adjoining pieces of lamellar bone (x 75)

in Paget's disease but carcinomatous metastases often lodge in them. On the other hand the tibia is but rarely the seat of metastases but may show a typical Paget disease lesion. If it is the lumbosacral spine and pelvis that are radiographically abnormal disseminated prostatic carcinoma may have to be considered. An increase in the serum level of acid phosphatase supports this latter diagnosis. The alkaline phosphatase is increased in Paget's disease and if the lesions are extensive, very high values up to and over 100 King Armstrong units may be determined. Such high values in the absence of liver disorders are virtually diagnostic of Paget's disease. The significance of only slight increases

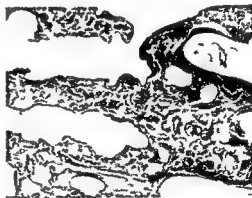
small proportion of sufferers who at anytime present clinical signs and symptoms are those with a few bones affected—a few vertebrae, one tibia perhaps, part of the pelvis one clavicle and the skull

There are no distinctive clinical features of the disease Aching pain in the affected part is the commonest symptom This is varied Sometimes it is pain



FIG 99—Section of bone in Paget's disease showing the characteristic mosaic pattern. Various shaped pieces of lamellar bone are joined to one another by darkly stained cement lines. Note the fibrous reaction still present in some marrow spaces (Haematoxylin and eosin $\times 18$)

FIG 100—Section of bone in late quiescent Paget's disease. The mosaic pattern is clearly shown by the unstained cement lines joining the irregular pieces of lamellar bone (Silver stain for reticulin fibres $\times 18$)



on movement or lifting sometimes pain on warming up in bed and sometimes in disease of the vertebrae a girdle sensation that Turner (1940) thinks may be due to a gradual narrowing of the intervertebral foramina. Enlargement and deformity of the vertebral laminae may occasionally lead to objective neurological signs from compression of the spinal cord. Thirty examples of this have been collected by Colclough (1949). The condition is fully described by Turner (1940) who points out that the upper level of compression in Paget's disease is nearly always at the second–fourth thoracic vertebrae and that since it is not the advanced case that presents spinal compression the neurological symptoms may first announce the disease. A special type of deformity with its own neurological consequences is platybasia caused by telescoping the cervical spine into the soft diseased bone of the base of the skull. Fracture of vertebrae affected by Paget's disease may result from rather trivial injuries. Sarcoma is a rather rare but important complication of the disease from which the vertebrae are not exempt

CHAPTER 5

OSTEOPOROSIS

REGINALD NASSIM

INTRODUCTION

THE whole concept of metabolic bone disease has been put on a firm scientific basis within the last fifteen years mainly through the work of Albright and his associates. Before that time it is true that demineralizing processes affecting the spine had been recognized and described (Meulengracht 1939, Black, Ghormley and Camp 1941, Burrows and Graham, 1945) but in all these papers there was only speculation regarding the aetiology although it was noted that the condition was commoner in the elderly and in women. However no true distinction such as is recognized today was made between osteomalacia and osteoporosis.

The approach to the whole problem of diagnosis, prognosis and treatment has been immeasurably simplified by improved biochemical techniques and by the intelligent use of metabolic balance studies. In this way osteoporosis and post-menopausal osteoporosis were clearly defined by Albright, Smith and Richardson (1941).

It is, therefore, depressing to record that the whole subject of osteoporosis still receives such scant recognition in British medical textbooks—a fact pointed out by Cooke (1955) in his admirable survey. This state of affairs is indefensible because post-menopausal and senile osteoporosis are the most common of all spinal diseases and are likely to increase still further in frequency with the steadily rising age of the population. From the economic point of view alone and apart from the symptomatic relief to the sufferers, awareness and early recognition of the condition would seem desirable. Intermittent courses of hormone therapy would simply and effectively treat many of the patients who at present attend their practitioners and are referred to overworked physiotherapy departments.

DEFINITION

There has hitherto been much confusion in the precise diagnosis of the condition because the term osteoporosis has so often been a radiological one—hence any demineralizing process of the spine and pelvis has been labelled osteoporosis.

There are many metabolic conditions that may give rise to an identical radiological picture and it is therefore essential to make a precise diagnosis and then to qualify it—for example osteoporosis due to the post-menopausal state or to Cushing's disease.

Correctly, osteoporosis means atrophy of bone from depression of osteoblastic activity. This allows normal catabolism to outstrip anabolism so that eventually there is insufficient osteoid tissue capable of normal calcification and hence a

in serum alkaline phosphatase may, however, be hard to assess. On the one hand small solitary foci of Paget's disease may put out no great amount of phosphatase, and on the other hand a moderate increase of alkaline phosphatase in the serum may accompany carcinomatosis if the metastatic deposits are osteoplastic. Sclerosing or osteoplastic metastatic tumours in bone may be of prostatic origin as is well known, but they also occur, and by no means rarely in disseminated cancers from breast, bronchus and other organs.

Conclusion

The cause of Paget's disease is quite unknown. Even the nosological category to which it should belong is unsettled. Its inclusion in this chapter on degenerative diseases may perhaps be justified only by the convenience of discussing it in this place.

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Immobilization

Immobilization is a well recognized cause for the rapid onset of osteoporosis. This is due to the removal of the normal stimulus to osteoblastic activity, namely the stresses and strains of weight bearing and active movement. When the spine or a limb is immobilized in a plaster cast there is an immediate rise in the urinary output of calcium which produces a negative calcium balance and the state continues till freedom is restored. This form of osteoporosis is commonly seen in patients immobilized for tuberculous disease of the spine. A similar picture occurs in patients with widespread paralysis, for instance severe anterior poliomyelitis. Apart from these conditions, long standing ankylosing spondylitis sometimes presents a severe degree of spinal porosis presumably caused by a combination of the inflammatory process and immobility of the spine.

Age

Osteoporosis tends to become commoner and more pronounced with advancing age as part of the general atrophy of tissue, such as affects also skin, muscle and hair. This process in the ageing bones is attributable to a combination of factors, the most important of which are (1) the diminished mobility of old people, (2) their tendency to a decreasing intake of food, and (3) changes in hormonal balance.

Menopausal osteoporosis

Women are not only affected more often than men but generally younger. The frequency of osteoporosis increases after the menopause and at an age usually before 65 years that cannot be attributed to senility. The spine may be seriously affected without evidence of atrophy of other tissues. This condition, called post menopausal osteoporosis, was defined by Albright, Bloomberg and Smith (1940) and Albright, Smith and Richardson (1941).

These workers had noted the physiological experiments demonstrating the effects of oestrogens and androgens on the skeletons of birds and mammals which have been summarized by Gardner and Pfeiffer (1943). They carried out balance studies on post menopausal women with osteoporosis and these showed that the women were in negative calcium and phosphorus balance and that both oestrogens and androgens had a beneficial effect on the calcium and phosphorus balance, causing a reduction in both urinary and faecal calcium. Androgens in addition favourably influenced the nitrogen balance and the two hormones appeared to act synergistically.

The state of the spine is the same in both senile and post menopausal osteoporosis and treatment with oestrogens and androgens is beneficial in both conditions.

Cushing's syndrome

Osteoporosis affecting the spine and pelvis has been well recognized in Cushing's syndrome for many years. Once again we are indebted to Albright (1942-43) for a rational interpretation. He postulated that the condition was due to excessive production of the adrenal S hormone which inhibited protein synthesis and was in contrast to the state in the adrenogenital syndrome where there was an excessive production of the N hormone which had androgenic effects. His predictions have been amply confirmed since the chemical

less dense radiographic image. There is no abnormality in the metabolism of calcium and phosphorus, and therefore no change in the serum levels of these constituents.

There are many different causes or combinations of causes that can lead to depression of normal osteoid formation and hence to osteoporosis.

Replacement of osteoid tissue

Certain broad principles may be accepted for the formation and normal replacement of adequate osteoid tissue.

(1) The genetic background of the individual must be normal. An abnormality that tends to the formation of abnormal connective tissue may prevent the formation of osteoid tissue as for instance in osteogenesis imperfecta.

(2) For the proper formation and replacement of osteoid a reasonably adequate diet must be provided with sufficiency of protein, vitamins and minerals.

(3) The most important stimulus to bone formation is adequate weight bearing and the physiological stresses and strains that go with it. Therefore any prolonged immobilization or paralysis will tend to diminish this normal stimulus to osteoblastic activity and lead to osteoporosis.

(4) It has become abundantly evident in the last decade that a properly balanced hormone state is essential.

The glands known to influence the skeleton are the pituitary, ovaries and testis, the suprarenal cortex and the thyroid and parathyroid glands.

With these basic facts in mind we can now discuss the various clinical conditions in which osteoporosis of the spine is probable.

CONDITIONS IN WHICH OSTEOPOROSIS MAY BE EXPECTED

Malnutrition

Severe and long continued malnutrition will eventually lead to atrophy of bone and in Western Europe it is unlikely to occur except in unusual circumstances such as famine during periods of war. It may, however, occur in countries where the standard of living is low and where the metabolism is adjusted to a low total calorie intake and the diet is deficient particularly in protein, calcium and phosphorus when the skeleton is light compared with that of people on an adequate intake as judged by western standards. The only cases of osteoporosis due to malnutrition seen by the writer were in patients with long standing anorexia nervosa. The condition may sometimes follow total or subtotal gastrectomy. Recently Atkinson, Nordin and Sherlock (1956) have described the skeletal changes in intestinal malabsorption due to long standing biliary obstruction and jaundice. The histological picture was mixed with evidence of osteomalacia in some osteoporosis in others or a combination of the two conditions in the same patient.

Deficiency of vitamin C

Vitamin C is essential for normal connective tissue formation and its absence from the diet gives rise to the well recognized skeletal changes that can occur in infancy. In the adult these changes are not usually seen in pure form but associated with multiple dietary deficiencies. On theoretical grounds deficiency should lead to porosis of bone.

SYMPTOMATOLOGY

Pronounced osteoporosis of the spine may be symptomless. The commonest presenting symptom is pain in the back usually confined to the lower thoracic and lumbar spine. Occasionally the pain is also of girdle type radiating around the rib margins and sometimes it may spread into the buttocks and thighs.

The onset is usually gradual though sometimes it is sudden with the feeling of something giving in the back, as after a jolt from stepping heavily off a kerb or bus. The ache is usually relieved by lying down though sitting or standing in one position for any length of time tends to make it worse as also does walking and any movement that may jar the spine—coughing, sneezing or straining. Some patients notice loss of height or a protuberant pouch from approximation of the lower ribs to the iliac crests.

In the elderly particularly the picture may be complicated by other degenerative conditions such as cervical spondylosis, lumbar disc degeneration and osteoarthritis of the hip.

DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

The diagnosis of the cause of the osteoporosis rests in the first place on an adequate clinical history and thorough clinical examination the value of which cannot be over emphasized, secondly on the radiological and biochemical investigations and thirdly in some of the less clear cut cases on a full haematological examination with bone marrow biopsy. In certain instances bone biopsy may also be necessary.

Assessment on the Clinical History and Examination

The important points to be considered are as follows:

The age of the patient, the duration of symptoms and whether or not immobilization has been a feature are significant. Information should also be sought about the diet and about bowel habits, because steatorrhea is a phasic disease and can be mild and without the passage of typical fatty stools. In both sexes evidence of normal sexual development should be sought and in females the time of onset of menstruation and of the menopause whether natural or artificially produced. It is also important to ask about previous operations particularly on stomach, pelvis or urinary tract, a history of renal colic or lithotomy may betray hyperparathyroidism.

The physical examination in most cases will reveal the presence of glandular abnormality with the well known stigmata of hyperthyroidism, Cushing's disease or acromegaly. Examination of the spine will usually show a smooth kyphosis or kyphoscoliosis. A sharp angulation suggests not a generalized spinal affection like osteoporosis but a disease affecting a part of the spine such as fracture, secondary metastatic deposits, Pott's disease or solitary or multiple myeloma. When loss of height has occurred through multiple vertebral collapse the lower ribs approach the iliac crests with consequent abdominal protuberance.

Measurements should be made of the span and of the distance from vertex to pubis and from pubis to heel. This will confirm the loss of height in the spine.

isolation of cortisone and hydrocortisone, the continued administration of which can produce spinal porosis and fractures

The distribution of the porosis in Cushing's syndrome resembles that in senile and post menopausal porosis but as Dent (1955) has observed the fractures which may occur in ribs or pelvis heal with the production of excess callus in contrast to its paucity in the senile and post menopausal cases

The pathological features have been described by Follis (1951) and Sissons (1956)

Thyroid gland diseases

Osteoporosis may occur in association with thyrotoxicosis but is not usually pronounced. The serum levels of calcium phosphorus and alkaline phosphatase are normal but there is an increased output of calcium in the urine and faeces. The negative calcium balance in this condition was demonstrated by Aub and his colleagues (1929). They showed also that the calcium excretion is decreased in myxoedema. The osteoporosis of thyrotoxicosis has been attributed to the negative nitrogen balance produced by the increased metabolic rate and general loss of tissue.

The histological picture in bone appears to be a mixed one. Follis (1953) found areas of osteoporosis and other areas with increased vascularity and osteoclastic erosions and in 6 of the 20 specimens studied osteoblastic borders were also noted.

Acromegaly

Osteoporosis may also occur in acromegaly. The cause is not known but is probably a combination of factors (Albright and Reifenstein 1948). These include depression of ovarian and testicular function which is known to occur, over activity of the thyroid or parathyroid glands which sometimes occurs, and possibly the high protein requirements in this disease.

Ovarian and testicular maldevelopment

Because oestrogens and androgens are necessary for normal skeletal development and because osteoporosis is common after the menopause and in old age it is not surprising that osteoporosis occurs in the absence of ovarian and testicular development. It has been noted by Albright Smith and Fraser (1942) in ovarian agenesis and there are corresponding findings in males.

Idiopathic osteoporosis

Finally there is a distressing group of cases in which severe osteoporosis occurs without known cause. The condition is called idiopathic osteoporosis and the cause may not necessarily be the same in all cases. The patients more often female than male tend to present in the second and third decades of life. There appears to be no endocrine abnormality. The course and symptoms fluctuate and treatment is usually disappointing. Nordin and Roper (1955) have described four women in whom osteoporosis and compression fractures were found during or soon after pregnancy and who improved with treatment. It is likely that porosis antedated pregnancy. No history of dietary deficiencies which might possibly have contributed was mentioned. The authors suggested that the cause might have been an excess of adrenal corticoids which is known to occur during pregnancy but as Cooke (1955) has pointed out the oestrogens also are increased in the blood during this period. Without further studies it is difficult to place these cases satisfactorily in any group.

DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

disc giving rise to the so called codfish vertebra. Grade III is a further extension of this with single or multiple vertebral collapse, as exemplified in Fig 102

The important points in the radiological diagnosis are

(1) Uniformity of demineralization. If only one or two vertebrae are affected or collapsed and the adjacent ones appear normal this is much more in favour of a localized disease or malignant deposit

(2) The presence in the ribs and pelvis of cysts such as occur in hyperparathyroidism or the multiple rounded translucencies of multiple myeloma or the



FIG 103 —Cushing's syndrome with fracture in pelvis showing excess callus

symmetrical pseudo fractures of osteomalacia which are so commonly found in the rami of the pubis and ischium and also in the ribs. In post menopausal and senile osteoporosis apart from the spinal deformity pathological fractures of ribs and pelvis are uncommon but they are less so in the idiopathic variety and in Cushing's syndrome. As Dent (1955) has pointed out there is a tendency for excessive callus formation in the fractures associated with Cushing's syndrome. This is illustrated in Fig 103 which shows the pelvis of a woman with Cushing's syndrome on whom subsequent operation was successful.

In post menopausal and senile osteoporosis there is usually little change in the long bones and skull and often none. This is not necessarily true of the idiopathic

and is useful in following progress. These measurements will also reveal cases of eunuchoidism in which the limbs are disproportionately long in comparison with the trunk. In rickets and osteomalacia the loss of height is due mainly to bowing of the lower limbs.

With this clinical assessment a presumptive diagnosis can usually be made and confirmed by the other diagnostic aids.

Radiological Diagnosis

The radiographic appearances of the spine are non specific and common to all conditions of demineralization of the skeleton.

It is of course true that radiology of other bones including the skull may be of prime importance in differentiating such conditions as hyperparathyroidism, osteomalacia and multiple myeloma.



FIG 102 —Typical porosis with codfish vertebrae

The typical appearances of the spine are well known and for convenience have been assessed in three grades (Donaldson and Nassim 1954). In Grade I porosis there is a coarsening and paucity of bone trabeculation with a stencilling of the outline of the vertebral bodies. Grade II proceeds to a swelling of the intervertebral

disc giving rise to the so called codfish vertebra Grade III is a further extension of this with single or multiple vertebral collapse as exemplified in Fig 102

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In post menopausal and senile osteoporosis there is usually little change in the long bones and skull and often none This is not necessarily true of the idiopathic

variety Fig 104 shows radiographs of the feet of a woman aged 28 years with marked demineralization and fractures of the metatarsals Her development and menstrual history were normal, and no biochemical abnormality could be found In any doubtful case a complete skeletal survey is necessary

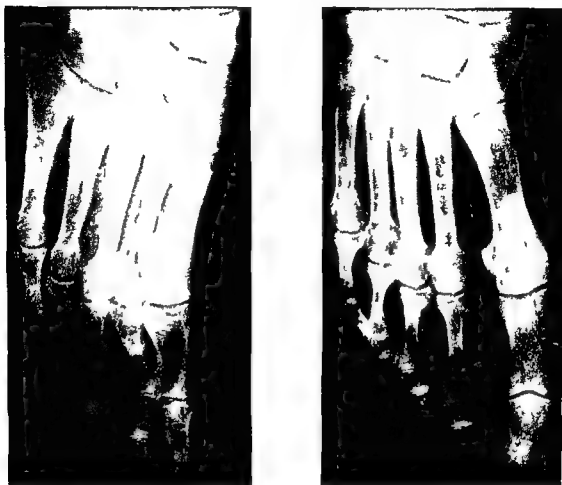


FIG 104—Radiographs of a foot in idiopathic osteoporosis showing fractures of metatarsals

Biochemical Diagnosis

Although the biochemical findings in the serum are normal in osteoporosis nevertheless these negative findings are essential in confirmation of the diagnosis and can help greatly in the differential diagnosis

Every patient with osteoporosis should have an estimation performed of serum calcium phosphorus alkaline phosphatase proteins and albumin globulin ratio

The urine should be examined for the presence of protein normal or abnormal and in doubtful cases estimations of the 24 hour urinary output of calcium should be checked If protein is increased in the serum further diagnostic help can be obtained by electrophoresis both of the serum and of the urine also if necessary after concentration

The important points in the differential diagnosis are

- (1) The serum calcium in osteoporosis is always within normal limits
- (2) The serum phosphorus is normal or, as has been noted by Albright Smith and Richardson (1941) and confirmed by Donaldson and Nissim (1954) on the high side of normal
- (3) The alkaline phosphatase is always normal
- (4) The urinary calcium may be high or normal depending on the duration of the condition. If the patient is for any reason immobilized the urinary calcium will tend to be raised. In the Metabolic Unit of the Royal National Orthopaedic Hospital and the Institute of Orthopaedics an excretion of more than 250 milligrams of calcium in the urine in 24 hours is regarded as abnormal.

In uncomplicated cases of osteoporosis the urinary calcium is not usually found to be as high as in hyperparathyroidism, multiple myelomatosis, Cushing's syndrome or thyrotoxicosis.

Any doubtful or abnormal values must be checked and other causes for the demineralizing process be considered. For instance, if osteomalacia is present the serum will be likely to show a low normal or low serum calcium and a low phosphate figure in the presence of a raised alkaline phosphatase and with low or absent urinary calcium. In hyperparathyroidism the serum calcium will be raised, phosphate low and phosphatase raised with a much raised urinary calcium. Difficulty may arise if renal damage has occurred. In this case the phosphate may be raised by retention and the calcium lowered. Under these circumstances the blood urea also will be raised and there may be acidosis. In multiple myelomatosis the serum calcium is usually normal but may be raised. However the serum phosphorus and alkaline phosphatase are normal. There is of course other evidence in the form of hyperglobulinaemia with high sedimentation rate and protein may or may not give the reactions of Bence Jones proteose. In Cushing's disease the serum calcium is normal and the urinary calcium output is high. The diagnosis depends on its well known clinical pattern and should cause no real difficulty except in mild cases. In the osteoporosis of thyrotoxicosis the serum figures for calcium, phosphorus and alkaline phosphatase are likewise normal and the urinary calcium is high. In acromegaly the serum phosphate is raised but calcium and alkaline phosphatase are normal. Once again most cases will be diagnosed without difficulty.

Blood Examination

The haematologist may be of great help in the differential diagnosis of osteoporosis. For in uncomplicated post menopausal or senile osteoporosis the blood count and sedimentation rate should be within normal limits. If anaemia is present and there is a raised sedimentation rate then these must be accounted for. The most common cause of osteoporosis with anaemia and a raised sedimentation rate is probably multiple myelomatosis or malignancy and the diagnosis can be confirmed by the finding of the typical cells by marrow puncture. In most cases there is also an increase in total serum proteins with typical elevation of the gamma globulin fractions which can be confirmed by electrophoresis.

Leukaemias occasionally give rise to osteoporosis affecting the spine and here again the haematologist may be able to give the correct diagnosis. Examination

of the blood may also help to confirm the diagnosis of Cushing's disease in which there is usually a high normal red cell count in the presence of a lymphopenia relative leucocytosis and absence of eosinophils

Rheumatic conditions such as ankylosing spondylitis or rheumatoid arthritis which are often associated with anaemia and raised sedimentation rate should give rise to no difficulty in diagnosis

Bone Biopsy

This procedure will be necessary only in a few obscure cases. The histologist can confirm the presence of osteoporosis by the presence of thin lace like and fragmented trabeculae in the specimen

Osteomalacia is recognized by its osteoid seams and hyperparathyroidism by the fibrosis and pronounced osteoclastic and osteoblastic activity. Cystic changes may also be revealed

Although the histologist can confirm the presence of osteoporosis he can give no indication of its cause. In recent years needle biopsy of vertebrae has been carried out but of this the writer has had no experience

The above diagnostic points are summarized in the Table

TREATMENT

The treatment naturally depends on an accurate diagnosis of the underlying condition. By far the largest number of patients are in the post menopausal or senile group. In these cases the administration of hormones leads usually to marked relief within a month

It is best to give a combination of oestrogen and androgen. The dosage need not be high. The writer has found for some years now that a combination of stilboestrol 1 milligram in the morning followed by a sublingual tablet of methyl testosterone 5 or 10 milligrams at night has been quite satisfactory

With this dosage the side reactions—such as bleeding, excessive vaginal discharge, tenderness of the breasts, acne or hoarseness of the voice—have been insignificant. The treatment is given in repeated courses lasting 2-3 months with breaks in between about three times a year. Apart from the symptomatic relief the effectiveness of the treatment may be judged by a drop in the serum phosphate. This change is probably produced by a suppression of anterior pituitary activity and a consequent increased clearance of phosphate by the kidney (Nassim, Saville and Mulligan 1956). Together with the hormone therapy an adequate intake of protein and calcium must be maintained. Diet does not usually play a part in post menopausal cases but it may well do so in some cases of senile osteoporosis

In those patients who give a history of inadequate intake of protein, calcium and vitamin D supplements may be necessary. Adequate calcium intake can best be achieved by drinking more milk

Immobilization should be discouraged as it promotes further porosis. Mechanical supports are usually unnecessary but on occasion they may help and then they should be of the lightest construction. Treatment for ovarian or testicular agenesis will be as recommended above. For the treatment of the porosis of Cushing's syndrome whether due to adrenal tumour or hyperplasia adrenalectomy either total or subtotal is the method of choice with subsequent

TABLE
FINDINGS IN OSTEOPOROSIS

	Serum calcium	Serum phosphate	Alkaline phosphatase	Urinary calcium	Blood	Sedimentation rate	Serum protein
Senile osteoporosis	Normal	Normal	Normal	Raised or normal	Normal	Normal	Normal
Post menopausal osteoporosis	Normal	Normal	Normal	Raised	Normal	Normal	Normal
Idiopathic osteoporosis	Normal	Normal	Normal	Raised	Normal	Normal	Normal
Cushing's disease	Normal	Normal	Normal	Raised	Normal* or increased cells	Normal	Normal
Acromegaly	Normal	High	Normal	Normal or raised	Normal	Normal	Normal
Thyrotoxicosis	Normal	Normal	Normal	Raised	Normal	Normal	Normal
Hyperparathyroidism	High	Low	Raised	Raised	Normal	Normal	Normal
Osteomalacia	Normal or low	Low	Raised	Low	Normal or anaemic	Normal	Normal or low
Multiple myeloma	Normal or high	Normal	Normal	Raised (protein also present)	Anaemic	High	High
Bone metastases	Normal or raised	Normal	Normal or raised	Raised	Anaemic	Raised	Normal

* Polycythaemia - leucocytosis

hormonal replacement. Similarly, for those cases arising in thyrotoxicosis the usual lines of treatment either medically with antithyroid drugs or by surgical removal of the gland, is adequate. The porosis of acromegaly can be helped by the administration of oestrogens and androgens as previously mentioned.

Although hormonal treatment produces marked symptomatic relief in most patients, and although balance studies show that the negative calcium balance can be corrected the amount of calcium retained daily is small and radiological improvement is disappointing even after 3-4 years of treatment.

The management of patients with idiopathic osteoporosis has been usually very disappointing. This group is probably not homogeneous. Most do not react favourably to the administration of hormones though occasionally both symptomatic and metabolic improvement can occur.

In some cases infusions of plasma or albumin have led to a positive calcium balance mainly by decreasing the urinary output of calcium (Albright and Reifenstein 1948, Anderson 1954) but the infusions have to be repeated every few months.

CONCLUSION

In conclusion although there have been encouraging advances in our knowledge of osteoporosis that knowledge is still very imperfect and many fundamental questions await an answer for instance why so many persons escape this particular facet of the ageing process and why fractures heal so readily in spite of severe porosis. Much further work is also necessary to understand the mechanism of the rapid porosis that follows immobilization in plaster and to discover methods to counteract it successfully.

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CHAPTER 6

TUBERCULOUS DISEASE OF THE SPINE

J A CHOLMELEY

CLINICAL PICTURE

THE COMMONEST regions of the spine to be affected by tuberculous disease are the lower thoracic and upper lumbar. In the individual vertebral body the disease usually starts anteriorly at or near the upper or lower margin. Two or more adjacent vertebral bodies are commonly affected and the nutrition of the intervertebral disc is interfered with. Consequently the disc becomes thinned. This narrowing of the intervertebral space is one of the earliest changes seen in radiographs of the tuberculous spine.

The accompanying inflammatory endarteritis causes cellular death of varying intensity in the vertebral body as a result of which pus formation occurs and sometimes bony sequestration, usually amounting to a coarse sand but sometimes leading to the formation of definite sequestra which may be recognized in the radiographs. In addition to bony sequestra a degenerate intervertebral disc may become separated.

In the thoracic spine paravertebral shadows, usually indicative of pus, are almost always seen in antero-posterior radiographs in the active stage of the disease. In the lumbar region this pus usually spreads along the sheath of the psoas muscle, where it can be seen as a distension in radiographs and often felt as a fluctuant mass in the corresponding iliac fossa. As healing occurs the pus in these abscesses thickens and may ultimately become calcified. The abscess shadow, however, may remain unaltered for a very long time, consisting largely of a thick wall.

In many cases activity of tuberculous spinal disease is not difficult to assess. The patient, child or adult, is obviously unwell, has lost weight, has night sweats and runs an evening temperature. The spine is stiff and painful with a localized kyphotic deformity which is tender on percussion; also there is spasm of the vertebral muscles. In addition to the above an abscess may be found on clinical examination. It is however not uncommon for several of these symptoms and signs to be absent in cases of proven active tuberculous disease. For example, the patient may neither look nor feel ill, there may be no evening rise of temperature, little pain and no obvious deformity. Radiographs may show little abnormality and tomographs may be needed to demonstrate clearly the site and extent of the disease.

It may in fact be impossible in the relatively chronic low grade type of tuberculous infection to decide when the active stage of the disease has ended. Clinical examination may show absence of pain and muscle spasm, the blood sedimentation rate may be normal, yet serial radiographs taken at regular intervals of, for instance, 4 months may reveal slow progressive destruction of bone.

Tomographs may be essential in some cases This slow progressive destruction may even occur after a successful spine graft when the efficiency of the graft may conceal some of the usual signs and may prevent collapse and bone apposition

CONSERVATIVE TREATMENT

The basic principle of treatment is rest—general rest both mental and physical, and local rest by some form of splintage For the tuberculous spine many forms of splints have been designed and used, and examples are the Bradford frame, Jones spinal frame plaster bed and the Berck box Treatment with hyperextension at the site of disease separates the diseased rarefied vertebral bodies during the active stage treatment with small pads placed above and below the kyphos encourages apposition of the diseased vertebral bodies throughout treatment and at the same time produces small compensatory curves above and below the lesion But probably all that is required is rest in a splint frame or plaster bed so designed that collapse at the site of disease is minimized Nursing the patient particularly a child in the prone extended position was advocated by Rollier This keeps the posterior spinal musculature in good tone acting as Rollier described it 'as a splint' This position can at least be used with advantage towards the latter part of the active stage of the disease

Antibiotics

Whatever form of splintage is used it should be combined with specific antibiotics such as streptomycin para amino salicylic acid (P A S) isonicotinic acid hydrazide (I N A H) and ortho hydroxybenzal isonicotinyl hydrazone (O H I H) Opinions vary on the combinations of these and other antibiotics that are most useful but a common and satisfactory combination is streptomycin 1 gramme daily P A S 15 grammes daily, divided into six equal doses and I N A H 200 milligrams daily divided into four equal doses These are average doses for adults For children under 10 years half these doses are used and for those between 10 and 16 years three quarter doses are used in each case if the children are of average weight for their age O H I H 200 milligrams daily divided into four equal doses can be used in place of P A S but should always be combined with pyridoxine 50 milligrams daily to prevent neuritis In all cases toxic complications must be watched for thus streptomycin may cause deafness and giddiness P A S may cause digestive upsets or rashes and O H I H, neuritis It should also be remembered that some people are abnormally sensitive to streptomycin and those administering the drug may develop rashes which may not only be very resistant to treatment but also recur whenever there is contact with the drug It is therefore advisable for those administering it often to wear rubber gloves or at least to wash their hands very thoroughly afterwards

The combination of streptomycin P A S and I N A H should be given for 3 months the case should then be generally reviewed and the same combination of drugs repeated for a further 3 months but with the streptomycin given only three times a week There is a tendency amongst those treating pulmonary tuberculosis to continue these or similar antibiotics for longer periods even up to 1 year and it may be considered desirable to do so in orthopaedic tuberculous lesions Cases of tuberculous disease have been seen where further lesions have

CONSERVATIVE TREATMENT

occurred a year or more after the original one in spite of large doses and even prolonged courses of antibiotics. If these prolonged courses are necessary it is all the more essential to be aware of the possible toxic complications and to watch for them because their onset is often insidious and may continue after the causative drug has been discontinued.

Value of antibiotics

It is very doubtful if any of the present antibiotics will eradicate tuberculous infection in a patient even if the drug is administered in adequate dosage for a long time. These antibiotics combined with routine conservative treatment do seem, however, to accelerate the rate of healing particularly in the more acute type of case, and in synovial joints to produce a higher percentage of safe mobile joints.

Unfortunately the natural healing process includes a localized endarteritis and fibrosis so that the more chronic and commoner type of lesion is not so favourably or quickly influenced by the antibiotics which have to reach the site of disease through the blood stream. This is particularly evident in spinal disease.

SURGICAL TREATMENT

Before the introduction of antibiotics with a specific action on the tubercle bacillus direct operations on tuberculous bone and joint foci before the disease was quiescent were generally avoided. However careful the operator sinus formation was a not uncommon complication this was always attended by the risk of secondary infection which might spread to the underlying bone focus. Consequently a secondarily infected tuberculous bone lesion developed that is a combination of a pyogenic and tuberculous osteomyelitis. Frequently such lesions never healed and the patient was left with a permanently or intermittently discharging sinus and the risk of amyloid disease. With specific antituberculous and other antibiotics, direct surgical approach to tuberculous foci is very much safer even in the active stage of the disease. Thus evacuation of soft tissue and bone abscesses and removal of tuberculous granulation tissue and sequestra can be accomplished with little risk of breaking down of the wound or secondary infection.

The posterior spinal graft for tuberculous disease of the spine has been reserved by most British orthopaedic surgeons exclusively as an internal stabilizing splint when the disease is quiescent. They have been unable to accept the teaching of some American orthopaedic surgeons such as Albee and Hibbs that such a grafting operation accelerates healing of the spinal lesion.

Operation in the active stage

Recently many orthopaedic surgeons in both America and Great Britain have been carrying out a direct attack on the disease in the vertebral bodies in the active stage. Macrae (1957) in treating thoracic spinal tuberculosis performs a bilateral costectomy, evacuates any pus and irrigates the area with streptomycin from each side through a catheter. He has found that healing and bony fusion between the affected vertebral bodies occurs in a surprisingly short time, such as three months. Wilkinson (1955) favours a more direct attack on the bone lesion. In the thoracic region he carries out a two rib costotransversectomy and if necessary, curettes and incises the vertebral bodies to remove granulation tissue.

and sequestra. In the lumbar region through a lateral approach he removes appropriate transverse processes thus entering the vertebral bodies; he then removes pus, granulation tissue and any sequestra that may be present. If there is then any cavity of appreciable size he packs this with cancellous bone chips thoroughly mixed with streptomycin powder. As a result of this procedure Wilkinson has reported acceleration of healing of the disease and more efficient and more frequent bony fusion between the affected vertebral bodies.

Conclusion

Through the discovery of specific tuberculous antibiotics direct surgical attack can now be carried out in the same way as in pyogenic non tuberculous osteomyelitis and arthritis. However, in the present state of knowledge it is still essential to continue conservative treatment of these tuberculous lesions though for a shorter period than before the advent of streptomycin. The evolution and healing of tuberculous lesions is slow and unlike those of the commoner pyogenic infections so that the final assessment of direct surgical attack on the diseased bone or joint focus will not be possible for several years, because prolonged and careful follow up will first be necessary.

TREATMENT OF COMPLICATIONS

Abscesses can be treated by aspiration but more extensive operations can now be carried out with safety under antibiotic protection. Thorough evacuation and resuturing of tuberculous abscesses is no longer followed commonly by sinuses and some writers have advocated and carried out successfully without sinus formation the excision of deep seated tuberculous abscesses such as the psoas abscess (Weinberg 1957).

Paraplegia

In tuberculous disease of the spine paraplegia occurs in about 10 per cent of cases and most often when the lesion is in the thoracic spine. Conservative treatment should first be tried and operation generally withheld for about 6 weeks and even then it should not be carried out unless there is motor loss. Exceptions are made if the paraplegia occurs and is increasing in depth in the course of conservative treatment if the course of the paraplegia is unusually rapid or if there is complete loss of motor power for a month. There are two operations of choice. If the paraplegia occurs early in the disease and more particularly in a child costotransversectomy should be performed as the compression is most likely to be due to thin pus. In all other cases and if costotransversectomy has not brought about improvement of the paraplegia within six weeks an antero-lateral decompression should be performed (Griffiths Seddon and Roaf 1956). In this operation caseous abscesses, sequestra or bony ridges can be located and as a rule adequately removed. Laminectomy should not be carried out for the relief of paraplegia in tuberculous disease of the spine because it is impossible to obtain adequate exposure of the vertebral bodies without risk of injury to the dura mater and the very nature of the operation must further weaken a spine which has already lost much of its stability by reason of the disease of the vertebral bodies.

The decision when to operate in Pott's paraplegia and what operative procedure

is best for each case is still a difficult one to make, and the writer considers that he cannot do better than to quote from Griffiths Seddon and Roaf (1956)

The majority of patients with Pott's paraplegia on admission to hospital show no absolute indication for operation. Motor power in such patients is usually severely impaired but by no means completely lost; sensation is moderately impaired or even normal, both splinters are working and spasms are few or absent. If the radiographs show a well marked paravertebral shadow and if bony disease has been present for less than 2 years, it will be wise to evacuate the abscess by costotransversectomy. This may, incidentally, cure the paraplegia. Apart from this consideration we would advise that the paraplegia should be given 6 weeks' full conservative treatment. If at the end of that time it is worse or no better decompression should be performed.

The patient with paraplegia beginning more than 2 years after the first symptom of spinal disease has less to expect from surgery than one whose paralysis arises early. Unhappily these late onset cases do not respond well to conservative treatment either. Immobilization and full chemotherapy should always be tried for if the paralysis is associated with continued activity of the bony disease or with reactivation of it such measures may well succeed. Most patients with such paralysis will require surgical treatment and some will be found very well suited to decompression as the cause of the paralysis is not always a bony ridge nor are all bony ridges irremovable.

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CHAPTER 7

NON-TUBERCULOUS INFECTIONS OF THE SPINE

J A MANTLE

INFECTING ORGANISMS

PROBABLY ANY organism pathogenic for man can settle in the vertebral column organisms already identified and reported include the staphylococcus streptococcus pneumococcus meningococcus, gonococcus *Micrococcus tetragenus* *Bacillus coli*, *Pseudomonas pyocyanea* *proteus vulgaris*, the diphtheria and diphtheroid bacilli the typhoid and paratyphoid salmonellae, the dysentery bacilli, the brucellae the streptothryces sporotrichon the coccidioides *Cryptococcus neoformans* *Treponema pallidum* and *T. pertenue* and the echinococcus [The haemolytic *Staphylococcus aureus* is the usual organism to be found] Despite their variety however non tuberculous infections are at present less common than tuberculous infections of the spine

PYOGENIC OSTEITIS OF THE SPINE

Large series of pyogenic infections of the spine have been analysed by Wilensky (1929) Kulowski (1936) and Guri (1946)

Aetiology

Pyogenic vertebral osteitis can occur at any age but it appears mainly in children between the ages of 10 and 15 years and is considerably more common in the male The lesions are almost invariably metastatic and quite often are preceded by an obvious infective focus elsewhere such as a furuncle styte whitlow quinsy, otitis media mastoiditis or osteomyelitis A history of recent injury to the spine is sometimes obtained Direct infection may follow wounds lumbar puncture or extension from an adjacent infective focus

Henson and Coventry (1956) and others have drawn attention to the urinary tract as a source of secondary spinal infections and it is probable that haematogenous spread occurs sometimes along the prostatic plexus of veins to the vertebral veins in the manner described by Batson (1940) The author has seen multiple infective foci in the pelvis and vertebral bodies after prostatectomy and periprostatic infection the distribution of the lesions being strongly suggestive of spread along this route

Pathology

In young patients the whole of a vertebral body is often involved whereas in older patients the lesions tend to be more discrete and involvement of spines pedicles or transverse processes is common The lumbar vertebrae are most often affected then the thoracic and cervical vertebrae the sacrum is least often affected

The lesion may be a rarefying osteitis a single destructive focus an epiphyseal separation or a subperiosteal lesion denuding the cortex and promoting the

formation of sequestra. Thromboangitis may also lead to sequestrum formation or decreased blood supply of the whole vertebral body. A Brodie's abscess may form from a localized infection of a vertebral body, the abscess cavity being surrounded by marked sclerosis. The proteolytic enzymes of the pyogenic bacteria can destroy the intervertebral disc substance, enabling the infection to spread across the disc to the neighbouring vertebral body. Early new bone formation usually leads to fusion of the affected vertebral bodies in such a case.

Marked collapse of the bodies with gibbus formation is uncommon in pyogenic vertebral osteitis. Martin (1946) considered that this is because the severity of the symptoms drives the patient to bed at an early stage of the disease, but it may also be because early sclerosis limits the destruction of the body.

An abscess starting anteriorly under the psoas muscle may lead to a retropharyngeal, mediastinal or retroperitoneal abscess. A lateral abscess may follow the psoas sheath or the course of a rib. A posterior abscess usually leads to pachymeningitis or dural compression. Pus from the lower lumbar bodies often collects in the hollow of the sacrum.

A tuberculous spine may of course become secondarily infected with pyogenic organisms if a tuberculous abscess ruptures through the skin or into the respiratory or alimentary tract. Seldman (1947) pointed out that conversely tuberculosis may also attack an area of spine devitalized by a pyogenic infection.

ACUTE OSTELITIS

Clinical course

The clinical course of pyogenic osteitis is usually acute in the younger patients and subacute or chronic in the older patients. The course appears to depend more upon acquired resistance than upon the virulence of the organism or the site of the lesion.

Acute osteitis may appear in one of several forms. The usual presentation is as an acute and fulminating disease of sudden onset with intense toxæmia. The toxæmia is generally a product more of the generalized bacteraemia than of the spinal lesion. The mortality has been high in the past, often because of complications. The acute form of the disease may subside into a subacute or chronic form.

A history of a primary infection elsewhere is commonly obtained. Spinal pain is severe; this is aggravated by movement, but unrelieved by recumbency and prevents sleep. There is local tenderness over the spine, and muscle spasm with rigidity or contracture of the spine. The high fever is often accompanied by rigors. At an early stage in the disease abscesses may be palpable, and signs of meningitis or paraplegia may appear.

Differential diagnosis

Pressure on the nerve roots with referred pain may simulate inflammation of other organs. Thus, involvement of the lower thoracic spine and nerve roots can simulate disease of an abdominal viscus such as the appendix, though the absence of real tenderness over the viscus, the absence of rigidity on abdominal palpation and the presence of tenderness over the spine should distinguish between the two conditions.

Referred pain to the hip with flexion deformity can simulate acute infection of the joint, though again there is tenderness over the spine and pain on attempting

CHAPTER 7

NON TUBERCULOUS INFECTIONS OF THE SPINE

J A MANTLE

INFECTING ORGANISMS

PROBABLY ANY organism pathogenic for man can settle in the vertebral column organisms already identified and reported include the staphylococcus streptococcus pneumococcus, meningococcus gonococcus *Micrococcus tetragenus* *Bacillus coli* *Pseudomonas pyocyanea* *proteus vulgaris*, the diphtheria and diphtheroid bacilli the typhoid and paratyphoid salmonellae the dysentery bacilli the brucellae the streptothryces sporotrichon the coccidioides *Cryptococcus neoformans* *Treponema pallidum* and *T. pertenue* and the echinococcus [The haemolytic *Staphylococcus aureus* is the usual organism to be found] Despite their variety however non tuberculous infections are at present less common than tuberculous infections of the spine

PYOGENIC OSTEITIS OF THE SPINE

Large series of pyogenic infections of the spine have been analysed by Wilensky (1929) Kulowski (1936) and Guri (1946)

Aetiology

Pyogenic vertebral osteitis can occur at any age, but it appears mainly in children between the ages of 10 and 15 years and is considerably more common in the male. The lesions are almost invariably metastatic and quite often are preceded by an obvious infective focus elsewhere such as a furuncle styne whitlow quinsy otitis media mastoiditis or osteomyelitis. A history of recent injury to the spine is sometimes obtained. Direct infection may follow wounds lumbar puncture or extension from an adjacent infective focus.

Henson and Coventry (1956) and others have drawn attention to the urinary tract as a source of secondary spinal infections and it is probable that haematogenous spread occurs sometimes along the prostatic plexus of veins to the vertebral veins in the manner described by Batson (1940). The author has seen multiple infective foci in the pelvis and vertebral bodies after prostatectomy and periprostatic infection the distribution of the lesions being strongly suggestive of spread along this route.

Pathology

In young patients the whole of a vertebral body is often involved whereas in older patients the lesions tend to be more discrete and involvement of spines pedicles or transverse processes is common. The lumbar vertebrae are most often affected then the thoracic and cervical vertebrae the sacrum is least often affected.

The lesion may be a rarefying osteitis a single destructive focus an epiphyseal separation or a subperiosteal lesion denuding the cortex and promoting the

PYOGENIC OSITITIS OF THE SPINE

massive, appear in about 2 months. Bony fusion between bodies or articular processes may be seen after 3 months. Dense sclerosis around a lesion of the body suggests the formation of a Brodie's abscess (Figs 107-108).

Differentiation between pyogenic and tuberculous spondylitis can be made radiographically but only by observing the later changes though a lesion in the neural arch or a process is less common in tuberculosis than in pyogenic osteitis.



FIG 105—Staphylococcal infection of the spine



FIG 106—Same spine as in Fig 105 3 years later



FIG 107—Brodie's abscess fourth lumbar body. One year after onset of pain



FIG 108—Abscess and backache still present (the patient had refused inpatient treatment) 4 years later. Same case as in Fig 107

Treatment

Immediate drainage of an extra dural abscess is imperative. laminectomy was formerly followed by wide bore tube drainage or repeated packing but now antibiotics have made primary closure permissible, as Blackburn and Jepson (1946) among others have shown.

spinal movement, and as Guri pointed out there is no pain on palpation of the posterior aspect of the hip joint or on percussion over the greater trochanter and extension is the only movement that is limited. Early meningeal involvement may simulate acute infectious meningitis, with the high fever and clinical signs of meningeal irritation.

Rheumatic fever, anterior poliomyelitis, peritonitis, pleurisy, pneumonia, typhoid fever, and transverse myelitis are among other conditions for which acute osteitis of the spine has been mistaken in the early stages.

SUBACUTE OR CHRONIC PYOGENIC OSTEITIS

Subacute or chronic pyogenic osteitis may follow an acute onset or the disease may be less acute from the start. Sometimes the onset is so insidious, and the course so slowly progressive, that the infection is overlooked for some months. There is little temperature elevation or other general reaction, but slight malaise and anorexia are usual. Limitation of movement of the spine and an area of localized tenderness to palpation or percussion are constant physical findings. Loss of weight and anaemia soon appear and can be severe.

Spinal pain is constant and may be severe, it is aggravated by movement and only slightly relieved by recumbency and it is often of a more intense nature at night. Pressure on a nerve root may mimic a prolapsed intervertebral disc with segmental pain.

In the chronic form an abscess may follow along fascial planes a long way from the spinal lesion and may resemble a cold abscess from a tuberculous spine.

Diagnosis

The main diagnostic difficulty is to distinguish chronic pyogenic osteitis of the spine from that of tuberculosis.

In both acute and chronic pyogenic osteitis, a polymorphonuclear leucocytosis and a raised erythrocyte sedimentation rate are almost invariable findings. Blood culture is positive if done early enough and urine culture is sometimes helpful. As Lack and Shelswell (1955) have shown, serological tests such as the staphylococcal anti-alpha haemolysin titration are occasionally helpful.

Needle biopsy or surgical exploration of a spinal lesion may be indicated when the diagnosis is in doubt.

Radiographic appearances

Radiographic evidence of the lesion appears only after a delay of 2 weeks or more: the earliest sign is a slight haziness and indistinctness of bone structure, this being followed by slight osteoporosis. In the patient who is severely ill it is not easy to obtain films of a sufficiently good quality to show these early changes. A fusiform paravertebral shadow may appear a little later, indicating an abscess. After a month or so porosis of the vertebral body may be almost complete in the younger patient and progressive compression of the body can follow. In older patients a more localized lesion is visible at this stage in the body or process of the vertebra, a destructive lesion of the body being accompanied by narrowing of the adjacent disc space. Sclerosis and new bone formation which is sometimes

massive appear in about 2 months. Bony fusion between bodies or articular processes may be seen after 3 months. Dense sclerosis around a lesion of the body suggests the formation of a Brodie's abscess (Figs 107, 108).

Differentiation between pyogenic and tuberculous spondylitis can be made radiographically but only by observing the later changes though a lesion in the neural arch or a process is less common in tuberculosis than in pyogenic osteitis.



FIG 105—Staphylococcal infection of the spine



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FIG 107—Brodie's abscess fourth lumbar body. One year after onset of pain



FIG 108—Abscess and backache still present (the patient had refused inpatient treatment) 4 years later. Same case as in Fig 107

Treatment

Immediate drainage of an extra dural abscess is imperative. laminectomy was formerly followed by wide bore tube drainage or repeated packing but now antibiotics have made primary closure permissible, as Blackburn and Jepson (1946) among others have shown.

Retropharyngeal abscesses can be drained through the neck and prevertebral and mediastinal abscesses should be drained by costo transversectomy. Psoas abscesses can be opened by a retroperitoneal approach with a lateral abdominal incision and presacral abscesses can be dealt with by an incision posterior to the rectum and facilitated by removal of the coccyx and, if necessary, resection of the lower portion of the sacrum.

The identification of the organism and the determination of its sensitivity to antibiotic drugs is necessary for definitive therapy. In many cases of acute spinal infection however the condition of the patient is so grave and the danger of delay so great that antibiotic treatment is started before the results of culture are available. Under such circumstances large doses of penicillin are appropriate since even if the organism be relatively insensitive it will probably be inhibited by high concentration of this drug.

Immobilization of the spine on a plaster bed should be continued until the general condition, the clinical signs, the erythrocyte sedimentation rate and the radiographic appearances show quiescence. If spontaneous fusion is not occurring posterior bone graft fusion may be considered. A pyogenic focus unlike a tuberculous lesion, rarely becomes active again but malalignment and instability can cause continued pain unless sound fusion is obtained.

The author has seen 3 patients with Brodie's abscess of a vertebral body in whom symptoms persisted for several years, and radiographs showed incomplete resolution after conservative treatment. If the symptoms are severe surgical drainage of the body is indicated in such cases.

Blood transfusion is often necessary for anaemia. Small doses of deep x ray therapy, short wave diathermy, infra red and ultra violet radiation have all been advocated in the treatment of the more chronic infections.

TYPHOID SPONDYLITIS

Typhoid infection of the bones becomes manifest some months after the enteric fever or during convalescence. occasionally it follows an unnoticed or subclinical initial infection. The spine and ribs are particularly singled out. Spinal lesions have followed both typhoid and paratyphoid fevers.

A low grade non suppurative periostitis or osteitis is the usual lesion but subperiosteal abscesses occasionally occur and the author has seen a large abscess in the iliac fossa associated with a spinal infection clinically indistinguishable from a tuberculous abscess but containing living paratyphoid bacilli. Ossification of the ligaments and fusion of articular facets is typical. fusion of vertebral bodies by bony bridges may occur.

Diagnosis

Pain is local but may also be referred along the nerve roots. There may be a history of recent typhoid fever but when the enteric stage of the disease has been subclinical diagnosis of the spinal infection is less easy. The pulse rate tends to be slow in relation to the pyrexia. Spinal rigidity and slight kyphosis are usual. Neurological changes with alteration of reflexes including an extensor plantar response can occur from the toxæmia without pressure on the spinal cord.

Blood, urine and stool culture should be performed. The Widal test shows

TYPHOID SPONDYLITIS

raised titre of agglutination for the responsible salmonella but recent inoculation as well as recent infection may give a similar effect. There is a leucopenia with decrease in the numbers of polymorphonuclear and eosinophil cells.

A positive Widal test or even a recent proved attack of typhoid fever does not necessarily mean that the spinal lesion is also typhoid. Lowered resistance, due to toxæmia and leucopenia is often followed by invasion with other bacteria particularly of the upper respiratory tract. These bacteria could themselves set up a metastatic vertebral infection.

Radiographic appearances

Radiographic appearances typical of typhoid show ossification of the ligaments, fusion of articular facets, areas of rarefaction in the bodies, narrowing of the disc space and fusion of the bodies. Several bodies may fuse, often at widely separated sites and fusion takes place more by bone bridges around the disc than by ossification in the disc. The bone bridge may be complete after 3 months but it will thicken considerably during the ensuing years.



FIG 109—Typhoid spondylitis (when first seen)



FIG 110—Same case as in Fig 109 8 months later

Treatment

Recumbency on a frame or plaster bed is necessary for about 2 months but operative fusion of the spine is not required.

Antibiotics such as Chloromycetin are effective against the typhoid group of salmonellae but their effect is less spectacular when bone lesions are established than in the acute stage of the disease.

BRUCELLAR SPONDYLITIS

Backache can occur early in undulant fever, but a true spinal lesion is not found until some months after the acute infection or during convalescence.

Brucella melitensis, *Br abortus* and *Br suis* affect goats, cattle and swine respectively but all three organisms may infect man. In Great Britain *Br abortus* is mainly responsible for the human cases and bone complications are less common here than in the United States of America or Southern Europe.

Lesions may involve vertebral bodies or epiphyses. Abscesses, if present are small occasionally sequestra are found.

Diagnosis

The symptoms include a history of recent fever which may not have been diagnosed as undulant fever pain which is often severe and rigidity of the spine. Deformity and palpable abscesses are rare. The course is more rapid than in tuberculosis of the spine. Paraplegia is rare though several early cases have been described (Jambon, 1950, Mantle 1955).

The temperature is intermittently elevated. The erythrocyte sedimentation rate is often high but is an unreliable test in this condition. Blood culture is often positive if done early enough. A positive agglutination test at a dilution of one in a hundred or more is significant but as Dalrymple Champneys (1950) has pointed out the agglutination may take up to 1 year to become positive or may even remain negative when the blood culture is positive. Negative agglutination zones also occur. When the agglutination test is positive a falling titre of agglutination is a better guide to progress than the erythrocyte sedimentation rate.

Radiographic appearances

Radiographic examination will show early and extensive bone proliferation and narrowing of the intervertebral discs, often with fusion of the bodies; rarefaction is absent.

Treatment

Good results in the treatment of uncomplicated undulant fever have been obtained with aureomycin, terramycin and Chloromycetin; some even better results have been claimed by combining aureomycin or terramycin with streptomycin. Once bone lesions are established however the efficacy of antibiotics is less certain and the author has seen continued progress of a brucellar spondylitis after a full course of antibiotics. Short wave diathermy and deep radiotherapy have been recommended. Immobilization in recumbency gives the best chance of achieving bony fusion but when spontaneous fusion appears unlikely operative fusion may hasten return to full activity.

SYPHILIS OF THE VERTEBRAL COLUMN

Lesions of the spinal column occur in congenital syphilis or in the secondary or tertiary stage of an acquired infection. The spirochaete reaches the spine by the blood stream or occasionally by direct extension of a gumma. Periostitis, arthritis, epiphysitis, osteitis and gumma may be found as well as neuropathic arthropathy secondary to neurological syphilis.

Much dense periosteal new bone forms about the periphery of the vertebral body. The arthritic form which often affects several joints presents hypertrophic lesions and excrescences on the bone with sometimes calcified material around the



FIG. 111 —Brucellosis of cervical spine on admission (By courtesy of Editor of *Journal of Bone and Joint Surgery*)



FIG. 112 —Same case as in Fig. 111. Condition 7 months later (By courtesy of Editor of *Journal of Bone and Joint Surgery*)



FIG. 113 —Brucellar spondylitis in lower dorsal spine (By courtesy of Editor of *Journal of Bone and Joint Surgery*)



FIG. 114 —Appearance 7 months later of case in Fig. 113 (By courtesy of Editor of *Journal of Bone and Joint Surgery*)

joints, disintegration of the joint with subluxation, may follow. Gumma usually affects only one body; it destroys the bone locally leading to slight alteration in the shape of the body and bony overgrowth at the vertebral margins, nerve roots may be infiltrated. There is no true abscess but material of a paste like consistency is sometimes found. Lesions are more common in the cervical spine than elsewhere in the vertebrae and a pharyngeal fistula may occur.

Clinical aspect

Clinically there is constant pain which cannot be relieved by either rest or plaster of Paris immobilization, the pain often radiates along the nerve root to one side only. In gumma of the spine there is less pain on movement, less limitation of movement and less muscle spasm than in tuberculosis.

Radiographic appearances

Radiographically the involved vertebra may appear larger and denser than its neighbours because of the formation of dense new bone around the periphery. Areas of osteoporosis may be seen with surrounding sclerosis. The discs are sometimes destroyed and bodies fused. New bone of great bulk and density may be seen about the articulations and as Brailsford (1953) pointed out this bone has a less clearly defined margin than is seen in osteophytosis of the spine. A scoliosis may be seen and lateral displacement may follow disorganization of the joints.

Serological reactions

The Wassermann reaction and the Kahn reaction are usually positive and the treponema immobilization test is invariably positive in a syphilitic lesion. The Wassermann reaction and colloidal gold reaction of the cerebrospinal fluid may be helpful. It is hardly necessary to point out that a positive serological test for syphilis indicating a past infection is occasionally found in association with a spinal lesion which is proved to be tuberculous.

Treatment

Treatment with potassium iodide and penicillin is effective but is sometimes supplemented with bismuth, mercury and arsenical preparations. Recumbency on a firm mattress may suffice but a plaster corset or Minerva jacket is necessary in the destructive forms.

YAWS

Granulomatous bone lesions resembling syphilitic lesions may be found after the secondary stage of yaws. These are common in the long bones but spinal lesions sometimes occur. Radiographs have shown multiple osteolytic lesions surrounded by sclerosis in the vertebral bodies. The Wassermann reaction is usually positive and this may lead to confusion with syphilis. The disease responds well to treatment with arsenical drugs and penicillin.

FUNGUS INFECTIONS OF THE VERTEBRAL COLUMN

ACTINOMYCOSIS

Actinomycosis is caused by the *Actinomyces bovis* and the organism reaches the spine by direct extension of lesions in the alimentary tract or the lung.

The lesions are granulomatous with progressive destruction of connective tissue, muscle and bone; after a time the mass breaks down being riddled with

abscesses, which often break through the skin to form sinuses. Usually several vertebral bodies are attacked and are honeycombed by abscesses. Ribs are often involved if the thoracic spine is affected.

Clinical aspects

The onset is insidious with spinal pain usually pain along the nerve roots and stiffness of the spine. Kyphosis is slight but infiltration and contracture of the spinal muscles on one side may give rise to a mild scoliosis. The mass may be palpable and the hard mass of an infiltrated psoas muscle is sometimes palpable abdominally. The discharge from sinuses and abscesses should be examined for the ray fungus and for sulphur granules at the first opportunity, as they are less likely to be found later.

Curettings and biopsy from sinus walls show (as well as chronic inflammatory cells fibroblasts and giant cells) lipoid cells these are stated to be characteristic. Paraplegia is rare although the meninges may become involved. Clinically actinomycosis may simulate tuberculosis or a sarcoma. The prognosis is poor and the diagnosis so difficult to make that in the past a high proportion of cases have been diagnosed correctly only after death.

Radiographic appearances

Radiographs show multiple areas of calcium loss mingled with areas of sclerosis. Typically the intervertebral discs are spared and there is no collapse. The ribs and transverse processes are often involved. A paravertebral abscess is often seen. The radiographic changes may also closely resemble those of tuberculosis.

Treatment

Radical excision is rarely practicable. Large doses of penicillin, combined with potassium iodide have often produced quiescence of the lesion. Isoniazid in combination with penicillin has been found helpful. The spine should be immobilized in recumbency in the early stages of treatment. Relapse after apparently successful treatment is common.

Streptothrix infections of spine other than actinomycosis have occasionally been reported. They do not appear to differ very greatly from infections with actinomycosis.

SPOROTRICHOSIS

Sporotrichosis is more common in the cutaneous form but occasional systemic and spinal lesions have been reported. The *Sporotrichum beurmanni* spreads along the lymphatics. Chronic granulomatous lesions with suppuration are formed, with sometimes a mass macroscopically resembling a gumma. The sporotrichum can be cultured from the pus, and the spore agglutination test of Widal and Abram gives agglutination at a high titre. Treatment resembles that for actinomycosis. The disease is chronic but rarely fatal.

BLASTOMYCOSIS

The term blastomycosis is properly applied to any infection with a yeast like organism. The *Cryptococcus gilchristi* is the species commonly found in the

cutaneous form which is far more common than the systemic form of blastomycosis. *Coccidioides immitis* is the species described in systemic blastomycosis. The organism spreads by way of the blood stream but direct invasion of the spine may follow a lesion in the lung.

The lesions consist of chronic granulomatous masses with suppuration. Microscopically there is tubercle formation with epithelioid cells and giant cells. The coccidioides are seen within the giant cells or in the tissues. Inoculation of pus into a male guinea pig produces a suppurative orchitis.

Radiographic appearances

Radiographs show foci of osteolysis with little reaction in the intact neighbouring bone. Lesions are usually multiple involving bodies, pedicles, laminae and transverse processes of the vertebrae and sometimes lesions of the ribs. The radiographic appearances however may resemble tuberculosis.

Treatment

Treatment with potassium iodide, penicillin and immobilization is like that for actinomycosis. Deep radiotherapy has been said to aid resolution. Before the introduction of penicillin, systemic blastomycosis was usually fatal.

SKELETAL CRYPTOCOCCOSIS

Skeletal cryptococcosis due to the *Cryptococcus neoformans* has been described by Gosling and Gilmer (1956). Lesions which may involve the spine are multiple, individually discrete and almost invariably osteolytic. Periosteal reaction may be present. Radiographically the appearances are like those of other forms of blastomycosis. Treatment with penicillin is assisted by thymol and iodides.

HYDATID DISEASE OF THE SPINE

The *Taenia echinococcus* may form hydatid cysts in the vertebrae or cysts formed in the adjacent tissues can involve the spine at a later date.

Clinical picture

The clinical progress is slow and there may be no symptom until the onset of paraplegia. Sometimes there is continuous backache which is usually not severe and sometimes pain along the distribution of one or more nerve roots.

Diagnosis

The intradermal test of Casoni and the complement fixation test using hydatid fluid as antigen are stated to be less commonly positive in osseous lesions than in generalized infestations. The white cell count shows eosinophilia.

Radiographic appearances

Radiographs show rarefaction of bone without reactionary limitation, the cortex may show evidence of erosion from outside with consequent change in the shape of the vertebra. Some calcification may be seen within the cyst and as Brailsford (1953) has pointed out the cyst may cause a large paravertebral shadow which can be mistaken for a paravertebral tuberculous abscess.

Treatment

Drugs have no effect upon hydatid cysts. Operative evacuation of the cysts with curdletting of its wall and injection of formalin is indicated if the cyst is accessible. Laminectomy may be necessary for decompression of the spinal cord, and it may have to be repeated a few years later for recurrence even though the cyst was satisfactorily evacuated.

CONCLUSION

The advent of the antibiotics has made the early diagnosis of vertebral infection more important than previously. In the past many non tuberculous lesions were regarded and treated as tuberculous until the shortened course of the disease and consideration of serial radiographs showing early bone formation and spontaneous fusion caused retrospective revision of the diagnosis.

When the treatment was not materially affected little harm ensued but today the patient will miss the benefit of modern therapy if the causal organism remains unidentified.

The history of the patient may provide valuable information about recent infections incurred before the onset of spinal symptoms but the clinical signs and early radiographs cannot be relied upon to give accurate differentiation between the various forms of subacute and chronic infective spondylitis. Every effort must be made to obtain a culture of the organism so that its sensitivity to the antibiotics can be determined instead of merely surmised.

Blood culture is positive in a high proportion of cases if performed early enough and it is suggested that blood culture should be performed in all cases of vertebral infection immediately they come under observation. The erythrocyte sedimentation rate and differential white cell count can conveniently be done at the same time though the information they afford is more inferential than particular. The various serological tests should be carried out otherwise many cases of syphilitic, brucellar, and other infections will continue to be treated as tuberculous.

Usually abscesses contain living organisms and the incision or aspiration of superficial abscesses is an easy matter. Paravertebral abscesses can be aspirated under radiographic control.

Needle biopsy of a lesion also done under radiographic control may produce a culture of bacteria as well as tissue for microscopical study. Early operation affords not only the opportunity for drainage of abscesses, but also material for culture and microscopical examination. Recent work has shown that even in tuberculous lesions early exploration is less hazardous than had been formerly supposed. The surgical approach to the vertebral bodies is becoming increasingly familiar and there is little doubt that in future a higher proportion of vertebral lesions will be explored.

There will remain a number of cases of spondylitis in which exploration is considered inadvisable and in which the various investigations have given negative or equivocal results. Under these circumstances it is permissible to use a combination of antibiotic drugs in the hope that the unknown organism will prove sensitive to one or the other. This course of action is reprehensible in many conditions but in infective spondylitis with no abscess or sinus even if the bacteria develop acquired resistance they will not threaten the community, they will remain incarcerated in the patient.

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CHAPTER 8

ANKYLOSING SPONDYLITIS

F DUDLEY HART

ANKYLOSING spondylitis, although closely related to rheumatoid arthritis, is considered in Great Britain to be a separate entity within the rheumatoid group of diseases. In the United States of America the two conditions are often included in the same series of cases and the individuality of ankylosing spondylitis is lost unless the case histories are recorded. Cases of ankylosing spondylitis do however, occur in which there is both spinal and peripheral arthritis but nearly always the presentation is distinctive enough to allow of a clear cut diagnosis. The intermediate group has been described as 'atypical spondylitis' (Sharp and Easson 1954).

AETIOLOGY

The aetiology of ankylosing spondylitis and the reasons for such facts as the predominance of the male sex and the usual onset of the disease between the ages of 15 and 35 years is still unknown. Certain old theories have recently been revived. Romanus (1953) for instance from a study of 117 male patients ascribed the condition to the dissemination of infection from the prostate and seminal vesicles and preferred the term 'pelvic spondylitis ossificans, secondary to genito urinary infection'. Ford (1953) reported 4 cases in which ankylosing spondylitis followed urethral infection in each case arthritis of the feet preceded spinal symptoms and signs. We have seen cases of this type developing slowly over a period of 4 years. Buckley (1948) stated that gonorrhoeal arthritis might attack spinal articulations, but that in such cases other joints were affected earlier. In most reported series the incidence of genito urinary infection corresponds to that seen in rheumatoid arthritis (Boland and Present 1945) or in the general young male population. The whole question however, of genito urinary infection is complicated by the patient's natural reticence in giving a full history. Lucherini and Cecchi (1952) considered that ankylosing spondylitis was not an infective inflammatory process though the initial stimulus might be infective in nature but a specific proliferative process.

Familial incidence

If a careful inquiry is made into the family history of patients with ankylosing spondylitis a definite familial tendency will often be found. Hersh and his colleagues (1950) observed the pattern of inheritance in 50 families and concluded that ankylosing spondylitis could be attributed to an autosomal dominant factor with about 70 per cent penetrance in the male and 10 per cent in the female. Ankylosing spondylitis has been reported in two pairs of fraternal twins (Campbell 1947, Rogoff and Freyberg 1949) and in sisters (Fraser 1950).

Mason (1951) reported an unusual family in which 3 certain and 3 likely members were affected and West (1949) found in his Bristol survey 9 families with more than 1 member affected. The familial incidence is also commented on by Blecourt (1951), Boni and Hautmann (1950) and Tegner and Lloyd (1949). Parr, White and Shipton (1951) recorded a significant family history in 11 per cent of their cases. It has been stated that if a female in a family is affected with ankylosing spondylitis there is always a male member similarly affected. While this in our own experience is untrue it is probable that many cases are missed or misdiagnosed.

Injury and climate

A not infrequent history of injury seldom proves relevant. Climate plays no important part. Ankylosing spondylitis is common for instance in warm dry climates such as that of Australia. Parr, White and Shipton (1951) stated that body build, climate, pre-existing tuberculous or gonorrhoeal infection and military service did not seem to play any aetiological role in their series of 100 cases.

INCIDENCE

Ankylosing spondylitis, a disease of military age, comes to the fore in war time; many cases first come to light during National Service. We have found that not infrequently the early symptoms appeared while the patient was at senior school. As in rheumatoid arthritis, early intermittent symptoms may precede the onset of the established disorder by several years; for example, intermittent swelling of a peripheral joint may occur once or many times before the steadily progressive and characteristic backache.

There are no certain figures as to the frequency of ankylosing spondylitis in the general population. West (1949) gave a figure of 1 in 2,000 (0.05 per cent). In the house to house surveys in Lancashire (Kellgren, Lawrence and Aitken Swan, 1953) and in Cambridgeshire (Jacobs, 1954) the incidence was negligible, only one case being revealed in over 3,000 people in the former and none at all in the latter.

PATHOLOGY

Ankylosing spondylitis is a disease characterized by a multiple arthritis leading to bony ankylosis of the posterior intervertebral, costovertebral and sacro iliac joints by ossification of the spinal ligaments and margins of the intervertebral discs. In many cases there is also rheumatoid arthritis of the proximal joints of the limbs. (Collins, 1949). Boland (1953) described the general pathological features as an inflammatory process in the sacro iliac, posterior intervertebral (apophyseal) and costovertebral articulations, the early changes resembling microscopically those seen in peripheral rheumatoid arthritis. He stated that the first changes seen are in the synovial membranes and consist of an infiltration of lymphocytes and plasma cells arranged in nests around the small subsynovial blood vessels; the synovial membranes proliferate (a villous synovitis) and at the same time some degree of joint effusion occurs. Gradually over a period of months or years granulation tissue grows out from the inflamed synovial membrane and invades the articular cartilage, eroding and destroying it, the joint space becoming filled with granulation tissue, the precursor of fibrous ankylosis and later bony ankylosis with disappearance of all vestiges of the joint.

Van Swary (1950-1951) studied the vertebral columns of 3 patients with ankylosing spondylitis, which was advanced in 2. The patients had died of myeloid leukaemia. He found a distinctive pathology unlike that of rheumatoid arthritis, the joint becoming obliterated by cartilage proliferation so marked that it passed across the joint space and invaded the cartilage on the opposite side, the old cartilage being reabsorbed by the new young cartilage. No cellular infiltration or vascularization was found. With the occurrence of cartilaginous fusion and the obliteration of the joint space the synovial membrane disappeared completely, having previously shown no proliferative changes. Calcium salts were deposited in the cartilage near the bone margin and in this heavily calcified area bone formed eventually replacing the cartilage.

Cruckshank (1951) obtained tissues from the diarthrodial joints in 12 cases from the hips in 6 cases and from the sternoclavicular joint, knee and sacro iliac joint in 1 case each. He found a process very like rheumatoid arthritis with subacute and chronic inflammation and hyperplasia of synovial tissue, destruction of articular cartilage by granulation tissue and later ankylosis usually bony. There was however a greater tendency to haemorrhage, a more marked and more frequent thickening of small vessels, usually in the form of endarteritis obliterans, and a greater tendency to bony fusion than in rheumatoid arthritis. He concluded that it was unjustifiable to classify ankylosing spondylitis as a variant of rheumatoid arthritis and he commented on the absence of nodules in spondylitis; nevertheless the picture was very like that of rheumatoid arthritis in every other way.

Collins (1949) considered ankylosing spondylitis a variant of rheumatoid arthritis and preferred the term 'rheumatoid spondylitis' as used in the United States of America.

It will be seen, therefore, that even morbid anatomists disagree about the relationship of these two diseases.

INVESTIGATIONS

Sheep cell agglutination test

Most workers have found that while the sheep cell agglutination test is usually positive in rheumatoid arthritis it is consistently negative in ankylosing spondylitis (Svartz and Schlossmann 1950). Ball (1952) for example found only 3 positive reactions in 203 cases of spondylitis (1.5 per cent) as compared with about 45 per cent positive in rheumatoid arthritis, since that time his incidence of positive reactions in rheumatoid arthritis has increased considerably. Our own figures are almost identical and our experiences similar. Foz Batalla and Barcelo (1951) found that in 59 cases of active rheumatoid arthritis 37.5 per cent gave positive results but in 25 cases of ankylosing spondylitis several with peripheral involvement only one doubtfully positive result was seen. Svartz (1953) in active and inactive cases of rheumatoid arthritis found that 78.5 per cent gave a positive reaction, whereas cases of ankylosing spondylitis were in general negative. This striking difference in the two diseases is of great interest and remains to be explained. There is some evidence however suggesting that the test is more often positive if rheumatoid nodules are present.

Sedimentation rate

Polley and Slocumb (1947) found normal rates in almost one fifth of their cases. We have been unimpressed by the value of the sedimentation rate as a guide to the progress of the disease or to prognosis because in some cases it remains normal in the face of acute symptoms and in others it remains elevated with the patient complaining of little or nothing. It is a useful diagnostic point when the patient first appears but in most cases it fluctuates at an elevated figure for many years.

SYMPTOMS AND SIGNS

Presenting symptoms

Few disorders give such characteristic histories and such classical physical signs as ankylosing spondylitis. Nevertheless though occasionally explosive in onset, it is more likely to be missed or incorrectly diagnosed in its early stages. In our own series initial diagnoses included fibrositis, malingering, anxiety states, prolapsed intervertebral disc, pyrexia of unknown origin and tuberculous disease of the spine, hip or shoulder.

In our series of 184 cases the initial symptom was as follows

<i>Symptom</i>	<i>Cases</i>
Low back ache or buttock ache or both	91
Low back stiffness without pain	18
Pain in hips, groins or thigh	16
Pain or swelling of ankles, heels or feet or both	13
Low back stiffness with slight pain	10
Sciatica	9
Pain or swelling of knees or both	7
Dorsal spine pain	6
Rheumatic fever	4
Pains in shoulders	3
Pains around the chest	3
Pyrexia of unknown origin with generalized aches and pains	2
Head going forwards	2

The initial symptom was therefore peripheral in 24 cases (13 per cent). The condition was misdiagnosed as rheumatic fever in 4 instances.

Peripheral joint involvement

Figures for peripheral joint involvement vary in different publications, perhaps partly because the duration of follow up also varies. Parr, White and Shipton (1951) noted a peripheral onset in 6 out of 100 cases. Kuhns (1951) in 6.5 per cent of 200 cases, and Hart (1954) in 24 of 184 cases (13 per cent). Forestier, Jacqueline and Rotes (1949) in a review of 200 cases of ankylosing spondylitis found that 78 per cent of 36 female and 56 per cent of 164 male patients had peripheral joint involvement at some stage of the disease. Sharp and Eason (1954) noted that 242 of their 332 cases were considered to be typical of ankylosing spondylitis, 59 were atypical and 31 were border line cases, the atypical group showing more peripheral joint involvement and a greater incidence (12 per cent) of the tendinitis and tendon nodules which are virtually non-existent in typical spondylitis.

SYMPTOMS AND SIGNS

Involvement of cervical spine

The cervical spine is invariably involved in advanced cases and often in relatively early ones though it is exceptional for it to be the site of the presenting symptom. Driving a bicycle or motor car may become difficult and painful, and the gradually developing forward stoop may be so insidious as to pass almost unnoticed by the patient for some months. Commonly the symptoms pass from lumbar region and buttock to the neck, apparently missing the dorsal spine, but less obvious symptoms referable to this area are usually elicited in a carefully taken history.

Acute onset

Ankylosing spondylitis may occasionally present in an acute, florid and extremely painful form. If the pain assumes an erratic course affecting the peripheral joints the patients are commonly thought to have rheumatic fever, particularly if under the age of 21 years. If the acute disease affects only the torso and girdle joints the correct diagnosis may not be made for some little time and a tabetic crisis may be suspected. In such cases the extreme discomfort is striking: the patient is in too much pain to be about but even bed affords no relief. These acute forms can be relieved effectively and fairly rapidly by cortisone, prednisone, corticotrophin or phenylbutazone (Butazolidin). Such florid cases are much less common than the insidious slow progressive ones where the diagnosis is often missed for some time. In civilian cases the diagnosis was made less readily than in service cases (Hart and his colleagues 1949): sometimes 3 or 4 years might elapse between the onset of symptoms and the establishment of the diagnosis.

Chest involvement

A characteristic and sometimes presenting feature of ankylosing spondylitis is the reduced rib excursion caused by fusion of rib on to the transverse process and body of the vertebra. The symptoms are usually stiffness of the chest wall, difficulty in fully expanding the chest and an aching and discomfort on deep breathing, coughing or sneezing. The reduction in costal respiration leads to over use of the diaphragm: a double exposure skiagram of the chest in inspiration and expiration will demonstrate little rib movement but a generous diaphragmatic excursion (Hart 1950; Hart, Bogdanovitch and Nichol 1950), the picture therefore differs from that seen in emphysema in which both ribs and diaphragm move little. In our series of 184 patients (Hart 1955) the initial chest expansion at nipple level when first seen by us was 1 inch or less in 88 cases; in only 34 cases was it over 2 inches.

Nodule formation

Many authors (for example Cruickshank 1951) have commented on the absence of nodule formation in ankylosing spondylitis. This though generally true, is not absolute.

Pain, swelling and tenderness of the manubriosternal joint is more common than in rheumatoid arthritis, but usually persists only for a few weeks. Tenderness over bony prominences may be a major complaint, the common sites being ischial tuberosities, pelvic brim, greater trochanters and occasionally the sacrum.

spinous processes sternum and ribs. Tenderness in the heel should always raise suspicion of ankylosing spondylitis especially in males between the ages of 18 and 30 years (Davis and Blair 1950)

The diagnostic importance of these tender areas in bone is obvious. They are often misdiagnosed commonly as tuberculous lesions, and the resulting therapy—prolonged immobilization—produces unfortunate results. It should be remembered that ankylosing spondylitis is a disease affecting bone as well as joint and ligament, it is an osteopathy as well as an arthritis and this aspect of the disease has often been overlooked.

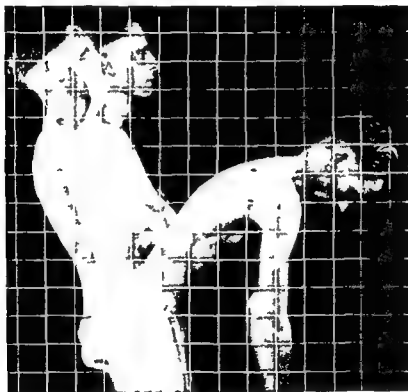


FIG 115—Triple exposure photograph showing the limited range of movement in ankylosing spondylitis. There is no alteration in the spinal contour.

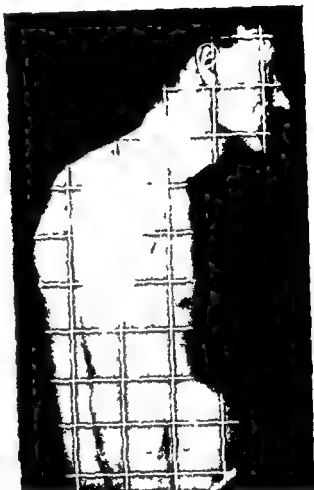
Physical signs

The classical physical signs are sufficiently well known and do not need emphasis. The 4 cardinal diagnostic pointers of stiffness of spine, reduced chest expansion, raised sedimentation rate and the classical radiological changes in the joints are usually all present. The lumbar spine is often flattened and is stiff as elicited by triple exposure photographs (Figs 115, 116). Head and neck movements are often similarly restricted, shoulders and hips less often. Bony tenderness may very occasionally be the outstanding finding. Tenderness of the ischial tuberosities may be present for many years and in 3 of our cases it was constantly present for

SYMPTOMS AND SIGNS

over 2 years though in most patients these symptoms abate after some months. These tender areas sometimes show radiological abnormality. Tenderness around the sacro iliac joints is exceptional and attempts to move these joints usually cause no discomfort.

FIG. 116.—Typical stance in fairly advanced ankylosing spondylitis. Dorsocervical kyphosis is often accompanied by a protuberant lower abdomen.



SPONDYLOMETRY

Measurement of spinal movement can only be done properly by using an instrument that eliminates hip and leg movement. The usual mode of assessing spinal flexion by measuring the distance of the fingers from the floor with the spine in full flexion is notoriously unreliable as it measures not only spinal flexion but also hip mobility, pain and tenderness in hamstrings and knee and several other factors. For the past 3 years we have therefore been using the 'spondylometer' designed by Dunham (1949) (Fig. 117). This instrument measures spinal movement between two points, those commonly selected being the sacrum and the vertebra prominens. Dunham has measured a number of normal subjects and compared them with patients suffering from ankylosing spondylitis and we have also measured the spinal range of movement in normal medical students. In our series the normal spinal range in young adults lay between 75 and 120 degrees and bore no relation

to the subject's ability or inability to touch the floor with his fingers. In ankylosing spondylitis these measurements are remarkably constant and provide the best measure of the progress of spinal stiffness. In early cases, in which pain rather than structural change restricts spinal movement, such movement is increased by treatment with cortisone, ACTH, or deep x rays, the response being much more rapid with the first two than with the last. Usually, however, there is considerable fixity of the spine, and, though the pain may be greatly relieved by different forms of treatment, the range of movement increases little.

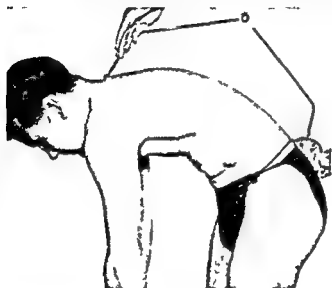


FIG 117—Measurement of spinal movement using spondylometer (By courtesy of Editor of the *Annals of the Rheumatic Diseases*)

ANKYLOSING SPONDYLITIS IN THE FEMALE

Though much less common in the female than in the male, ankylosing spondylitis may be more common than has been supposed in that sex (Simpson and Stevenson 1949, White 1953) because it is usually milder and therefore more readily overlooked. In a 20 year series Tyson, Thompson and Ragan (1953) had 60 female patients as compared with 450 male (1:7.5). The average age at onset in the female patients was 25 years. In most respects these authors found the disease differed little from that in males, though a greater proportion of female patients had involvement of the cervical spine and symphysis pubis. Four of our female patients have borne children, one by caesarean section and 3 normally (Hart, Bell and Organe 1951). There was no real aggravation of the condition nor was there deterioration in the vital capacity. On the other hand, there was no symptomatic improvement such as is commonly seen in patients with rheumatoid arthritis who are pregnant. White (1953) also found that no marked regression occurred during pregnancy and noted that the disease rarely gained impetus after confinement.

ASSOCIATED CONDITIONS AND COMPLICATIONS

Iritis and iridocyclitis

Iritis and iridocyclitis are relatively common in ankylosing spondylitis. Buckley (1948) gave 10 per cent as the approximate incidence; the association is more

ASSOCIATED CONDITIONS AND COMPLICATIONS

likely to be higher in long standing cases (Hart 1951) Iritis may occur during the course of the spondylitis or may precede it (Chen and Sun 1951) No case of scleromalacia perforans in a spondylitic has so far been published



FIG 118—Advanced ankylosing spondylitis in a woman. The figure on the left shows the spine in maximal flexion, the spine being completely fused. On the right the patient is seated on a chair she invented. On this she attends the theatre and cinema reasonably comfortably.

Cardiac complications

There has been emphasis in the last few years on cardiac findings at autopsy in cases of rheumatoid arthritis and it has also been held that aortic valvular lesions, particularly aortic incompetence, are not uncommon in ankylosing spondylitis. Bernstein (1951) examined 352 patients with ankylosing spondylitis, in 37 per cent of whom the disease had lasted for more than 10 years. Clinical evidence of heart disease was found in 43 per cent; 10 patients had valvular disease, 6 with aortic insufficiency. Electrocardiograms showed some abnormality in 47 of 190 patients, the most frequent change being prolonged conduction time in 29. She stated that the cardiac complications which arise in spondylarthritis ankylopoietica are of benign nature. Even in the case of valvular disorders, symptoms of insufficiency are not very prominent, even in advanced age.

Kuhns (1951) recorded 200 patients with ankylosing spondylitis observed during 5–20 years. Of these 29 had died, mostly from unrelated conditions, but 5 had died from rheumatic heart disease, and 2 from superadded endocarditis, presumably bacterial. He stated that the most serious hazard to life in rheumatoid spondylitis seemed to be a previous rheumatic involvement of the heart.

Intercurrent disease

Dyspepsia

Dyspepsia, with or without peptic ulceration is a real hazard among these predominantly male patients aged 20-40 years particularly as almost all current forms of treatment, whether it be aspirin, phenylbutazone (Butazolidin) cortisone corticotrophin, prednisolone or deep x ray may aggravate symptoms with the production of occasional melaena, haematemesis or even perforation. There is little statistical evidence that peptic ulceration is more common in patients with ankylosing spondylitis than in those of like age without this disorder, but it is common experience that it is frequent enough to constitute a real hazard in therapy. The dyspeptic spondylitic is extremely difficult to treat.

Pulmonary tuberculosis

Pulmonary tuberculosis occurs in 5-8 per cent of patients with ankylosing spondylitis. The importance of the co existence of these diseases lies in treatment for rest is the basis of treatment of one disorder graded activity and exercises of the other.

DIFFERENTIAL DIAGNOSIS

To be considered in the differential diagnosis are the following

Fatigue and postural strain, tuberculous spinal disease, osteoarthritis, hysteria, anxiety and other psychoneurotic states, injury, prolapsed intervertebral disc, cervical spondylosis, other rheumatic disorders such as Reiter's syndrome, rheumatoid arthritis, continued rheumatic fever, psoriatic arthropathy, metastatic malignant disease of the spine, spinal tumours, osteochondritis, spondylolisthesis, undulant fever or enteric fever, osteoporosis and osteomalacia, osteitis condensans ilii and fluorosis.

All the many causes of backache have in fact to be considered. The main difficulty lies in the early case, correct diagnosis may be virtually impossible without some months of careful observation. A most important differential point lies between tuberculous spinal disease and ankylosing spondylitis, prolonged immobilization of a spondylitic with an incorrect diagnosis of tuberculous disease may aggravate stiffness and induce ankylosis.

RADIOLOGY

The earliest radiological evidence of ankylosing spondylitis is usually ascribed to the sacro iliac joints. With experience of 500 cases Scott (1942) emphasized this and considered that sacro ilitis was the first demonstrable radiological lesion that every spondylitic had sacro iliac joint changes and that bony ankylosis of these joints always preceded spinal lesions. Golding (1936) reviewed 91 cases of ankylosing spondylitis and found sacro iliac changes in all of them. He believed that this was the earliest demonstrable manifestation. It is now generally held that for all practical diagnostic purposes the disease can be said to start in these joints.

Sacro iliac joints

The appearance of the normal joint varies considerably with the radiographic projection and in films taken for assessment of progress it is important to standardize projection and penetration as much as possible. If the incident ray strikes the

pelvic brim at nearly a right angle (as in the lordotic patient) the upper joint margins are seen and the joint is foreshortened. If the incident beam is directed at right angles to the body of the sacrum, there is no foreshortening and a greater length of articular margin is projected. Adequate radiographic penetration is most important as it is vital to demonstrate both the anterior and posterior margins of the joints. Underpenetrated and underexposed films are misleading.

The joint may be assessed on the usual standard projections but the joint space cannot be clearly seen unless it lies in the direction of the incident beam. This view is obtained by rotating the pelvis 15–20 degrees to the side away from the joint being radiographed. The ilium and sacrum are then viewed independently without the overlap seen on anterior posterior films caused by the obliquity of the joint and with a clear joint cavity between the two bones. This view of the sacro iliac joint is essential in assessing the extent of the pathological changes. Tomography also is useful in demonstrating the joint space at different depths.

The normal sacro iliac joint of the adolescent looks different from the adult joint. The subarticular bone cortex is not seen and the joint space is not delineated by sharp linear well defined cortical bone as in the adult. The joint space appears relatively wider and its bordering bone is finely irregular. This normal appearance of the adolescent joint must be appreciated because it bears many resemblances to the diseased adult joint. Rolleston (1947) emphasized this similarity and compared the earliest changes in ankylosing spondylitis with a patchy persistence of these adolescent features.

In our experience the earliest radiological changes are usually apparent in the lower half or third of the joint as seen in the antero posterior or oblique projection the normal clearcut joint space becoming blurred. Much of this loss of sharp definition is due to ilial sclerosis (Borak 1946), this sclerosis alone curving crescentically backwards will in an early case give the false impression of joint destruction and for this reason alone well centred oblique projections are essential. As the disease progresses and the joint fuses the sclerosis lessens and finally disappears. In the early stages a patchy zone of para articular porosis gives an apparent widening of the joint erosion being seen between joint space and adjacent sclerotic ilium. The bone texture presents a mottled appearance of alternating porosis and sclerosis the relative predominance of which varies from case to case. Sometimes juxta articular porosis precedes and is more pronounced than sclerosis.

With progressive cartilage destruction the joint space gradually disappears bony ankylosis occurring irregularly here and there. Trabeculae can be seen traversing the joint progressing until complete fusion occurs. The final state is often one of complete bony ankylosis with well defined trabeculae traversing somewhat porotic bone bony sclerosis having usually completely or largely disappeared. A 'ghost joint' can often be seen even in cases completely ankylosed scratchy linear markings showing the line of the obliterated joint space.

The changes described may run their course within months or years depending on the tempo of the disease as any one appearance may remain almost unchanged for many years during apparent quiescence. Almost without exception the changes are bilateral and nearly always symmetrical but occasionally especially in the early stages the disease may be considerably more advanced on one side.

ANKYLOSING SPONDYLITIS

Knutsson (1950 1953) followed up 99 patients who had the so called typical changes of ankylosing spondylitis in the sacro iliac joints and he suggested the following arbitrary grading of radiological change

(1) *Early changes* Blurring of the joint margins with iliac sclerosis and a normal joint space

(2) *Destructive stage* Loss of joint space erosions and increasing sclerosis

(3) *Ankylosing stage* Gradual bony ankylosis with loss of sclerosis

Knutsson considered that the radiological changes in the sacro iliac joints were diagnostic and specific and were found in no other disease with this opinion many disagree

Spine

Several pathological changes in the vertebrae and their associated ligaments are demonstrable radiographically

Vertebral synovial joints

The apophyseal joints may be an early site of the disease in the spine but the changes are not easy to demonstrate a series of coned views of the spine taken in different degrees of obliquity averaging 90 per cent for cervical 70 per cent for the thoracic and 35 per cent for the lumbar spine is required and even then not all of these apophyseal joint spaces may be demonstrated The results are often disappointing and the changes equivocal Tomography is an academic excursion rather than a helpful investigation as the diagnosis has already been made on radiological changes elsewhere

Vertebral bodies

The angles of the vertebra may become sharpened and the vertebral body becomes squared assuming the appearance of a child's brick Romanus and Yden (1952) studied serial films of the spine in 114 men with ankylosing spondylitis and found osteolytic lesions at the margins of the vertebral bodies later re ossifying with remodelling to the squared appearance

Generalized osteoporosis of the spine has often been described as an early and important feature of ankylosing spondylitis (Buckley 1948 Scott 1942) We have not been impressed by this except in patients confined to bed or in those with long standing disease who have undergone prolonged spinal fixation such as in a plaster bed Mowbray Latner and Middlemiss (1949) also considered that general spinal porosis was rare it is certainly not often seen in the ambulant male adult with active disease

Spinal ligaments

Ligamentous calcification and later ossification is first seen in the anterior and posterior longitudinal spinal ligaments The calcification may be difficult to demonstrate radiologically as only when the incident beam is tangential to a plaque of calcium is a radiographic shadow cast Collins (1949) demonstrated at necropsy that the calcification was initially in the outer laminae of the annulus fibrosus extending into these fibres from the epiphyseal rings of the bordering vertebrae Calcification and subsequent ossification of the annulus fibrosus and the longitudinal ligaments may continue until the typical picture of bamboo spine is

seen. There is loss of lumbar curve and a gradual continuous thoraco-cervical kyphosis, the whole of the vertebral column is then rigid and kyphotic. Ligaments also are also involved and the median interspinous ligaments ossify and may form a continuous median longitudinal bar of bone. If this is accompanied by ankylosis of the apophyseal joints and their capsules an appearance of 3 longitudinal parallel bars may be produced.

The disc spaces are usually unimpaired and are of normal height probably because of the splinting effect of the ossification. If the discs are degenerate before significant ossification occurs considerable disc narrowing may be seen.

Thorax

Both the central costovertebral joints and the lateral costotransverse joints are affected and early changes are extremely difficult to demonstrate but advanced changes show up readily on well penetrated antero posterior films the ribs appearing to spread out as they fuse with the transverse process and body (or bodies) of the vertebra.

The involvement of the costovertebral and costotransverse joints causes much rigidity of the thorax and loss of vital capacity (Hamilton 1949, Hart 1950, Hart, Bogdanovitch and Nichol 1950). The manubriosternal joint is commonly involved and less frequently the sternoclavicular joints. This further limits the expansion and elevation of the thoracic cage in inspiration. Apophyseal joint changes and ossification of the intervertebral ligaments are of less significance in this respect. A double radiographic exposure of the chest on a single film, in full expiration and in full inspiration shows in a normal subject considerable movement of both the ribs and the diaphragm during the full range of respiratory excursion. The patient with ankylosing spondylitis with thoracic involvement shows very little rib movement—sometimes none at all—whilst the diaphragmatic movement is of normal or increased range. In generalized emphysema there is both diminished rib and diaphragmatic movement.

Pelvis and hip joints

It is generally true that ankylosing spondylitis is a disease of the trunk and proximal joints the more peripheral the joint the less the likelihood of its permanent involvement.

The symphysis pubis is not uncommonly affected and may become ankylosed. Attention has been drawn already to bony tenderness as a common physical sign in ankylosing spondylitis the radiological concomitants of such lesions have also been reported. Guest and Jacobson (1951) described oval areas of porosis irregular bone margins with whiskery outgrowths into soft tissues and occasional bitten out areas in the pelvis on its outer aspect but not on its inner margin which is free from muscle insertions. These authors found that 74.4 per cent of the 90 patients studied had such lesions commonly in the ischial tuberosities and iliac crests. Extrapelvic lesions of this nature are not so readily recognized and when found in the vertebral body or humeral head may be mistaken for tuberculous lesions (Hart 1955).

Of the limb joints the hips are most likely to become radiologically involved and to cause severe disability. Complete bony ankylosis may occur. Scott (1942) found that 10 of 300 cases of ankylosing spondylitis were diagnosed as tuberculosis of the

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hip by both clinician and radiologist. Subsequent immobilization in plaster was followed by early and complete bony ankylosis of the hip with greatly increased disability. To avoid this error of diagnosis the pelvis should be radiographed and the sacro iliac joints assessed whenever an atrophic or infective arthritis of hip is suspected. Almost without exception it will be found that if spondylitis be the cause the sacro iliac joints are obviously involved and probably ankylosed.

TREATMENT

It has been customary in the past to emphasize the importance of a good spinal position. Patients were confined to bed and were often put in plaster cases which were gradually increased in extension so that the relief of pain might run parallel with fusion of the spine in a good position. Such treatment has now been completely superseded by a more active approach. There is no doubt that this therapeutic programme was productive of much pain and crippling, when the patient was released from his plaster he had in many cases not only a stiff fused back but stiff fused hips and a rigid thoracic cage also. In our own series those misdiagnosed as suffering from spinal tuberculosis and treated by immobility showed the worst end results: in most their hips were either largely or entirely ankylosed. In most patients attending our unit for the first time wearing spinal supports the vital capacity was diminished and full inspiration prevented in greater or lesser degree as a result of our findings we have entirely discontinued the use of spinal supports. Sharp and Easson (1954) adopted the same attitude.

Today patients with ankylosing spondylitis are advised to maintain full mobility short of severe spinal strain from the start. We would prefer a spondylitic to have an active occupation such as gardener or plumber rather than an indoor sedentary post for patients feel themselves to be better when active and much worse when kept at rest. Awakening in the morning is usually their worst time because after some hours of deep sleep they are stiff and in pain and may take from 2 to 3 hours to become fully mobile. Not uncommonly patients set an alarm clock to wake them at some hour in the night when they do a few simple exercises and then return to bed: in this way they are able to mobilize more rapidly in the morning and their working day is made easier. Other patients report that morning stiffness is reduced when their spines are kept gently agitated at night as in moving vehicles ships or aeroplanes. We therefore encourage the patient to undertake all physical activities that do not involve severe strain to the spine: cricket swimming golf and tennis usually prove beneficial if the disease is not too advanced. An active life with the normal overbreathing of simple exertion is better than set breathing exercises at set times though these are also beneficial. Spinal stiffness does not cripple a man but stiffness and pain in the hips does: indeed fitness for work often depends entirely on the presence or absence of hip involvement. A few weeks or months in bed or in a plaster shell may mark the beginning of progressive hip involvement.

Sharp and Easson (1954) recorded that in their group of typical spondylitics 19 had been at some time in the course of the disease immobilized in plaster beds or on frames for months or years. Of these 19 5 were virtual statues with rigid dorsal and lumbar spines and fixed or grossly limited hips and 2 of them had practically fixed knees. Only 6 similar cases were encountered among the remaining 223 patients in the group.

Physiotherapy

The most important item in physiotherapy is exercise the physiotherapist should teach and encourage postural and breathing exercises and simple bodily movements which the patient should do regularly at home or at work every day. When in hospital we prefer our patients to have the freedom of the building and its grounds and we encourage simple physical training short of severe strain. Though a large variety of different physical methods have been advocated none is very dramatic in effect though any method may on occasion appear helpful. Various forms of applied heat are as likely to aggravate as to ease symptoms in this disease. Treatment at a spa will often benefit the more advanced or partly crippled patient.

Analgesics

Aspirin

In any of the rheumatic disorders an adequate and regular analgesic regime is of primary importance. In milder cases only an occasional dose may be needed to relieve temporary pain but many patients need a regular hour to hour analgesic cover. Acetylsalicylic acid in any of its forms taken at set times may make all the difference to the patient's ability to cope with his daily work. The morning dose is usually the most helpful. 15 grains of aspirin, with a small glass of milk and a biscuit on waking will make the most difficult task of the day, getting up, less of a painful proceeding.

Phenylbutazone (Butazolidin)

This drug has proved its worth essentially as a long acting slowly excreted analgesic. The fact that it helps the spondylitic in all stages of his painful disease suggests that its benefit lies essentially in relief of pain. If the patient can tolerate phenylbutazone it is perhaps the most effective drug in the treatment of the advanced or early painful case. Daily doses of 200-400 milligrams are usually well tolerated by the male patient with advanced spondylitis but the therapeutic effect may be inadequate. Increasing the dose to 600-800 milligrams a day may produce ease but will often be followed by gastric intolerance. Other toxic side effects such as oedema, skin rashes, buccal ulceration and defects in blood clotting are more rare while leucopenia and agranulocytosis are rarer still. Gastric irritation is the most frequent and important hazard and may lead to haematemesis and melaena. Although there is no effect on stiffness when the disease has led to fixation, relief from pain enables the patient to do more with less discomfort. Toxic effects, if they occur usually do so within 6 weeks of starting treatment. There is considerable variation in individual response to the drug, some tolerating it extremely well others suffering side effects within a few days on small dosage. If the drug is effective it usually continues to give relief for a long time.

Cortisone, hydrocortisone, prednisone, prednisolone and corticotrophin

The improvement caused by these substances in the rheumatic disorders depends on various factors of which an important one is their anti-inflammatory or oedema-reducing action. In many cases of ankylosing spondylitis this factor is not of primary importance and though the patient may feel and look better the effect on function may not be dramatic. Other patients respond dramatically, obtain much

relief of pain and are able to do more with less discomfort. For prolonged maintenance therapy it is unusual to exceed 75 milligrams of cortisone or hydrocortisone or 20 milligrams of prednisone or prednisolone daily by mouth. Although these doses may be exceeded for short periods to cover acute exacerbations it is generally accepted as unwise to continue high dosage. The hazards are peptic ulceration sometimes with melaena, haematemesis or even perforation, any of the many clinical features of Cushing's syndrome, and masking of acute infections. It is easier to start steroid therapy than to stop it, and what may be started as short term therapy may run into a long term project as patient and doctor find that this treatment cannot be discontinued because of aggravation of symptoms on reducing the dose.

Corticotrophin (ACTH) either intramuscularly or intravenously is used in the treatment of an acute exacerbation. It may also be used to produce adrenocortical stimulation when cortisone or prednisone is being withdrawn.

Deep radiotherapy

Deep radiotherapy is the only form of treatment that produces an effect lasting longer than the therapy itself. While the beneficial effects of cortisone, corticotrophin and phenylbutazone last only a few days after withdrawal of the drug, deep radiotherapy frequently does not appear to produce amelioration in the clinical picture until towards the end or after the cessation of treatment, and benefit may last months or years.

Procedure

Richmond (1951) has summarized the ways in which deep radiotherapy has been given. The first is the wide field therapy which, though now given up, did much to focus attention on the value of irradiation in this disease. The second is what Richmond calls 'periodic conservative therapy' in which a relatively conservative dose of x rays is directed to the region of the spine from which symptoms appear to arise at the time. It is based on the consideration that ankylosing spondylitis is a relapsing remitting disease; further treatment may be given as symptoms arise elsewhere. The third method is the 'localized semi-intensive' method. In most instances therapy is directed to the sacro iliac joints and the lower lumbar spine in the first instance. Though local improvement in the site irradiated is the rule, recrudescence of pain at a higher level in the spine is common. The fourth method is the generalized semi-intensive method which is favoured by Richmond, by Windeyer and by McWhirter (1945) and many others in Great Britain. It consists of irradiation over the sacro iliac joints and entire spine irrespective of the stage and localization of the disease. A common dosage is 2 000r surface dose to each section of the spine, usually over a period of 4 weeks, employing x rays of 1.9 millimetre copper half value layer (250 kilovolts with 1 millimetre copper, 1 millimetre aluminium filtration). This means that 100r is given to the sacro iliacs, lumbo, thoracic and cervical regions daily for a total of 20 treatment days.

In our own unit we prefer to treat only the areas affected, though we include areas of very mild symptoms or signs, because experience has taught that relief of pain in one section of the spine is frequently followed by complaint of apparent exacerbation in previously less painful untreated areas. The dosage given varies from 1 000 to 1 250 skin dose, treatment being given only on alternate days to any

particular area. The routine procedure adopted (Hart and his colleagues 1949) was to use x rays of kilovoltage 180-200, filtration 1.5 millimetres copper, half value layer 1.7 millimetres of copper: the skin dosage in each field never receiving more than 150r in any one day. Treatment was not given to more than two fields daily: the period of therapy being spread over 3-4 weeks. The advantage of this smaller dosage and more localized application is that further courses can be given later and systemic upset is much less. With the generalized semi-intensive method using 2 000r total (skin) dosage further courses of therapy are not advisable. In addition to irradiation of the spine we have given deep radiotherapy in smaller dosage of a few hundred roentgens to painful and tender areas elsewhere: for example pelvic crests, knees and feet: often with very satisfactory results.

Comparative results

Desmarais (1953) compared the effect in ankylosing spondylitis, osteoarthritis and rheumatoid arthritis of radiotherapy applied to different areas in different dosages. Deep x ray therapy in ankylosing spondylitis gave infinitely better results than in osteoarthritis: the results in rheumatoid arthritis being the most disappointing of all: not uncommonly joints flared up in consequence of treatment.

In our experience most patients with ankylosing spondylitis obtain pronounced improvement in symptoms. This is not paralleled, except in very early cases, by marked improvement in range of spinal movements: relief from pain is more striking than relief from stiffness. Why deep x rays improve the patient is not known and it is debatable whether it alters the natural course of the disease, probably it does not. Whether deep x ray treatment can bring on natural remission is still open to question. It is, however, the one form of treatment which usually gives symptomatic relief for a considerable time.

Side effects

There are certain toxic side effects. Melaena with secondary anaemia occurs occasionally: particularly on dosages of 200r and upwards: rarely on lower dosage. Van Swaay (1950-1955) reported necropsy findings in irradiated patients who died of myeloid leukaemia: and Court Brown and Abbatt (1955) drew attention to this complication particularly in patients subjected to repeated courses of therapy. Aplastic anaemia is rare. Sharp and Easson (1954) reported a case. Activation of quiescent pulmonary tuberculosis and irradiation nephrosis are also possible hazards.

Surgery

The orthopaedic surgeon can help in preventing ankylosis in a bad position. It is important to remember that an anatomical position is not always the best from a functional point of view. The patient who can sit but not stand may be better off than the one whose capabilities are the other way round. Ordinary arthroplasties are most disappointing. For ankylosed hips excision of the femoral head and neck sometimes succeeds: but even with a stabilizing osteotomy does not allow unassisted walking: because stiffness prevents the actual flexion of the spine necessary to bring the centre of gravity over an unstable hip when it takes weight. Osteotomy osteoclasts of the spine which has been publicized in recent years is rarely justifiable and should be reserved for most carefully selected cases: for there is a very real risk of paraplegia.

PROGNOSIS

Accurate prognosis is difficult. Death has occurred from injury, the rigid fused neck of a spondylitic breaking like a long bone in such cases death may be instantaneous or occur after some weeks or months of paraplegia. Intercurrent infection often respiratory may cause death, tuberculosis was responsible in one case in our series. Amyloid disease is not uncommon in the late stages of any of the rheumatoid diseases and ankylosing spondylitis is no exception. Cardiac deaths as noted above are not uncommon later in life. Kuhns (1951) stated that life expectancy in the usual case of ankylosing spondylitis is the same as with other individuals with a similar amount of crippling, in our experience previous misapplied treatment has caused much of this crippling. In fitness for work and ability to carry on with his daily duties, the spondylitic is more happily placed than the rheumatoid the great majority of sufferers attending our unit are at full work and have been able to continue so for many years. Blumberg and Ragan (1956) found that of 121 patients followed up 76 per cent were working regularly and only 5 per cent were reduced to bed or chair existence.

SUMMARY

In order to study the disease more closely attempts are being made to differentiate it more strictly from other disorders. There is some evidence that typical and atypical forms react differently to treatment and that the atypical variety may include more than one disease process.

There is still disagreement about the essential pathology. Incrimination of genito urinary infection has been revised but not to the satisfaction of most workers. The aetiology remains as in rheumatoid disease in general unknown.

In therapy emphasis is towards activity and exercise when possible and away from unnecessary immobilization.

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CHAPTER 9

CERVICAL SPONDYLOSIS

VALENTINE LOGUE

THE PROLAPSED intervertebral disc in the form of a soft nuclear herniation has become a commonplace in diagnosis in the 23 years since its first description as a clinical entity. The symptoms and signs of its presence at each of the three levels of the spinal canal—lumbar, thoracic and cervical—have been described in every detail.

A different nosological entity which has been revealed by radiography and which is referred to as spinal osteoarthritis, osteophytosis, spondylitis and various other names has also been recognized for a long time as being an affection of the intervertebral discs along with other local structures, but its significance in the production of neurological disorders, as distinct from its own symptoms of local pain and stiffness, has only been discovered in recent years. That this condition, best referred to as spondylosis, may occur at any level in the spine is also well known from studies on the cadaver (Shore, 1953) and from radiological investigation of the spine (Boyle, 1954). It has been found by these methods that in middle age spondylosis is most common at the levels of the following intervertebral discs: C 5/6 and C 6/7, T 7-11 and L 3/4 and L 4/5. It is generally acknowledged that the incidence increases with age and over the age of 80 years all discs are affected at all levels in the spine. However, with regard to neurological complications it is found that these are uncommon at the thoracic and lumbar levels—in the lumbar region because the spondylosis is related to nerve roots and not to the spinal cord and roots are more adaptable and more resistant to pressure by the relatively small encroachments in the spinal canal that spondylosis produces. In the thoracic region the sparing of the cord is probably related to the comparative immobility of the spine and also to the small amount of posterior osteophytosis that occurs there.

It is in the cervical region that involvement of spinal cord and nerve root by spondylosis is commonest—for a number of reasons (see below) of which the mobility of the cervical vertebrae is probably the most important. Nervous complications of cervical spondylosis are probably much commoner than was formerly supposed. Pallis Jones and Spillane (1954) have shown in a series of 50 patients admitted to hospital for conditions independent of the spine that 75 per cent had cervical spondylotic changes visible radiographically and no less than 50 per cent of these showed abnormal neurological signs, although these were not necessarily all related to the spondylosis and many of the signs were slight—such as an extensor plantar response, reduction of vibration sense or an alteration of reflexes—and did not give rise to any disability.

However, in some instances the changes produced in the spinal cord and nerve roots are severe and cause progressive crippling, but why this small minority is destined to suffer serious neurological complications is still not known. Certainly the degree of neurological damage does not necessarily appear to be related to the severity of the radiographic changes.

PATHOLOGICAL CHANGES IN THE INTERVERTEBRAL DISCS

The essential feature of a spondylosis which produces damage to the spinal cord and nerve roots is the backward protrusion of the annulus fibrosus into the spinal canal for a varying distance. The projection takes the form of a narrow bar of fibrocartilage enclosed above and below by a bony spur or ridge emanating from the margins of the adjacent vertebrae and extending for part or the whole way across the spinal canal. This spur formation may be confined either to the posterior intraspinal aspect of the disc or to its anterior surface, or it may affect the whole of the circumferential margin of the disc.

Although occasionally the fibrocartilage may proliferate and form a small soft knob on the apex of the protrusion, it is the hard transverse ridge of fibrocartilage, bone and fibrous tissue that is the characteristic nosological entity implied by the term cervical spondylosis and with which this chapter is concerned. This annular protrusion is clearly quite different from the localized soft herniation that occurs classically in the lumbar region and that is also seen but less often in the cervical spine.

The protrusion of the annulus in cervical spondylosis is associated with narrowing of the disc seen radiologically, but on the other hand narrowing of the disc is not necessarily associated with a protrusion. For instance x ray examination of the cervical spine may show equal narrowing of several discs only one of which may show actual spur formation.

The precise pathological basis of this type of spondylotic protrusion is not clearly known. The basic change seems to be degeneration of the disc substance comprising both annulus fibrosus and nucleus pulposus. The nucleus shrinks and loses its resilient properties and consequently the vertebral bodies approximate. The annulus wherein hyalin changes and coarsening of the fibres are taking place as part of the degenerative process now bears the brunt of the stress of movement of the joint. As a consequence of the approximation of the vertebrae and the increased mobility between them the annulus tends to bulge out so that the periosteum to which it is attached tends to be lifted up with proliferation of the subjacent bone to form the osteophytes which grow out above and below enclosing the fibrocartilage of the annulus between them. With continuing movement the spurs tend to increase in height. As mentioned above there may occasionally be a soft cap on the protrusion from proliferation of the already protruded fibrocartilage.

The fundamental causes of the disc degeneration responsible for the annular protrusion are even more obscure. There are the natural processes of decay associated with ageing which begin earlier in the intervertebral discs than in other specialized tissues, appearing in the third decade. They are associated with a progressive reduction in the fluid content of the discs particularly that of the nucleus pulposus. This desiccation and increased firmness of the nucleus means loss of bulk and of resilience. In some people this degenerative process seems to be more severe at an earlier age than in others so that the stress of normal daily neck movements has a greater effect in predisposing to spondylotic changes. Why the change should affect some people earlier than others and why it should be confined to a single disc in about a quarter of the cases is not clear. Trauma has been adduced as a possible cause but the history of a single initiating injury is very rarely obtained although it is well known that violence to the cervical spine

already the seat of spondylosis may have a disproportionately serious effect. It is possible, however, that injury at an early age and long forgotten may be the basis for spondylosis later, and this may explain perhaps why only one disc may be affected and the other discs appear normal on radiography.

Certain occupational diseases seem to predispose to cervical spondylosis and congenital anomalies, such as fusion of the bodies of two or more vertebrae are often associated with spondylosis of the disc situated immediately above or below, presumably from excessive stress at these levels, but the possibility that the disc itself is congenitally mal developed has to be borne in mind.

Butler (1955) has suggested that osteochondritis in childhood may be the origin of spondylosis in adult life and certainly the radiographic appearances of fully developed cervical spondylosis resemble closely the changes seen in the thoracic spine in this condition.

SITE OF SPONDYLOSIS

Spondylosis can affect any one of the six cervical discs from that between C2 and C3 vertebrae to that between C7 and T1 with the highest incidence at C5/6 and C6/7. Usually more than one disc is affected and occasionally all six.

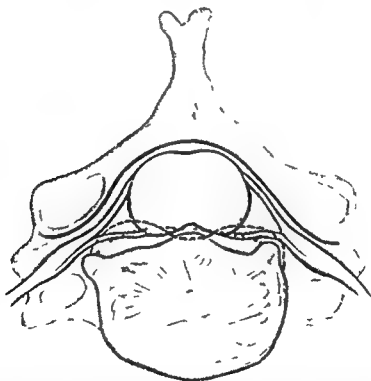


FIG. 119 —Semi diagrammatic drawing to show the various situations of annular protrusions — intraforaminal dorso lateral and the median protrusions which are indicated by dotted lines.

The precise situation of the osteophytic protrusion within the spinal canal is important because it determines whether the spinal cord or the nerve root or both is going to be affected. Fig 119 shows the various types of protrusion. A bulge in the midline will compress the whole spinal cord (and the anterior nerve roots

SITE OF SPONDYLOSIS

that emerge at this level) but a bulge just to one side of the midline causes a mainly unilateral cord compression with a Brown Sequard type of syndrome. More often a complete bony bar passing from one side of the canal to the other is seen which may affect not only the spinal cord but also the nerve roots on one or both sides. A further variety consists of a localized protrusion which arises from the lateral portion of the disc in relation to the nerve root and is either dorso lateral or intraforaminal in situation. In the latter site the protrusion may not be derived from the disc but from hypertrophic lipping of the margin of Luschka's (neuro central) joint, as Cave, Griffiths and Whiteley (1955) have emphasized. From the practical point of view the distinction is not important because treatment conservative or surgical will be the same in either case. Luschka's joints are para diarthrodial and are set on the lateral margins of the vertebrae facing into the intervertebral foramina. Osteoarthritic lipping of their margins usually appears in association with cervical spondylosis but it may occur as a separate entity in the presence of reasonably normal discs.

CLINICAL SYNDROMES

The clinical syndromes that may arise from the annular protrusion in each of these situations are therefore those of (1) root compression alone causing pain down the arm (brachialgia or brachial neuritis) as a result of dorso lateral or intraforaminal protrusions (2) compression of the spinal cord by median or paramedian protrusions or (3) a combination of both. In addition there will be symptoms in and about the neck itself, as a result of the local disease process in the spine.

Local symptoms

The symptoms referable to the neck are mild, in spite of severe spondylosis demonstrated by radiographs and it is usually symptoms of nervous system involvement that bring the patient to hospital. However occasionally there is a history of attacks, often recurring of slight aching or stiffness in the neck with perhaps some radiation into the shoulders, the mild symptoms being dismissed by the patients themselves as manifestation of rheumatism or fibrositis. The patient may sometimes notice restriction of head or neck movement but the neck symptoms are rarely obvious and are overshadowed by the symptoms of nerve root or spinal cord compression. In the developed case it is usually possible to demonstrate some reduction of mobility of the cervical spine mainly lateral flexion. Restriction of antero posterior flexion and extension tends to be obscured by the free movement of the head at the atlanto occipital articulation and even in the presence of florid neurological signs the cervical movements may sometimes be quite unrestricted.

NERVE ROOT COMPRESSION

Symptoms

The predominant feature of nerve root compression from a laterally placed protrusion is pain distributed according to the level of the disc lesion and the nerve root involved. With protrusion at a high level C 2/3 or C 3/4 pain is greatest in the scapula and shoulder. In lesions of the lower discs pain extends down the

length of the arm and into the fingers and often on to the chest. In fact it is now well known that compression of the C7 root on the left side may give rise to pain which radiates into the arm and chest, simulating very closely coronary artery thrombosis.

The pain has the usual persistent aching quality that is associated with nerve root compression at any level in the spine. It is commonly subacute typically occurring in an attack which lasts for a few weeks to perhaps 1 or 2 months. The attacks tend to recur the intervals of freedom becoming less, and they often finish up as a persistent intractable aching down the limb exacerbated by any activity and relieved to a variable extent by rest. Occasionally a really severe attack of pain occurs which resembles that produced by the less frequent soft nuclear prolapse though radiography may indicate spondylotic changes of many years standing such an acute incident is difficult to explain by an essentially chronic disease process. In these occasional severely painful attacks the neck movement may be considerably restricted by muscle spasm in contrast to the slight restriction in the usual case (*see above*). In general, however, the root symptoms are subacute or chronic permitting the patient to carry on work with mild restriction of activity. In addition to pain, paraesthesiae of the fingers is a common complaint.

Signs

Examination of the arm will disclose the neurological signs which are now well known and do not require a detailed description. They consist of, perhaps, slight selective wasting of muscles reduction or abolition of tendon reflexes which will vary according to the nerve root involved, and sometimes sensory change in an area of dermatome distribution.

Analysis of the pain

On analysis of its character and quality the pain in cervical nerve root compression it will be found to be of two types there is a deep boring, aching pain felt mainly in the more proximal muscles those of the neck scapula deltoid, biceps and pectorals and there is a sharper lancinating pain often occurring in shock like attacks radiating more peripherally in the limb often into the fingers where they are usually associated with paraesthesiae. The distribution of pain is widespread even with a single root irritation and in the very acute attacks patients may state that the whole arm is aching and they are unable to specify a particular area related to a single root distribution.

Mode of production of the pain

The mode of production of painful symptoms by root compression has been clarified considerably by the researches of Frykholm (1951). He carried out operations on patients with brachial neuritis under local analgesia and was able to stimulate the affected nerve roots and note the quality and distribution of the resulting pain and then abolish conduction by injecting the anterior and posterior rami with procaine. It was found that the deep proximal aching pain was reproduced exactly by mechanical stimulation of the compressed ventral (motor) ramus this pain could be abolished completely by procaine block of the related sensory (posterior) ramus alone the motor root being left intact stimulation of

the motor root would still produce muscle contraction, but not pain. The most reasonable interpretation of these observations was that the proximal aching pain arose from persistent spasm in groups of muscle fibres in consequence of motor root irritation, pain sensation from these fibres being relayed back to the central nervous system by the related posterior ramus.

When the sensory root alone was stimulated a sharp, shock like pain was felt which radiated not only widely into the proximal muscles but also distally in a more circumscribed fashion in the sclerotome distribution as well as into the dermatome area where paresthesiae were experienced. Frykholm was able to show in this way that in nerve root compression the painful symptoms were referred into the myotome, sclerotome and dermatome areas by irritation of both the motor and sensory roots, with a wide diffuse distribution proximally and a narrow and more localized radiation distally.

Segmental pattern of innervation

There has been in the past a tendency to regard the segmental pattern of innervation in the upper limbs as being rather fixed, with a small range of variation caused by pre fixation or post fixation of the nerve roots contributing to the brachial plexus. Frykholm has demonstrated however, that the variation is wider than has been thought and that the component nerves of the plexus may move two segments up or down, for instance the cutaneous supply of the little finger, usually regarded as being subserved by the eighth cervical nerve may in fact be innervated by the sixth sensory root. This has a practical application because it renders it unwise to operate for root compression with a localization based solely on clinical methods and makes myelographic confirmation of the level an essential preliminary to surgery.

Anatomy of the nerve root

The manner in which the nerve root is involved by laterally placed protrusions has also been investigated by Frykholm. In order to understand his explanation it is necessary to describe in some detail the anatomy of the nerve root as it emerges from the theca (Fig. 120). At the level at which the nerve root makes its exit the dural sac sends out a funnel shaped prolongation called the root (axillary) pouch with a curved upper and lower border so that the nerve root is permitted to run a gently curving course out of the theca, thus avoiding a sharp bend at its point of emergence. At the apex of the root pouch there are two openings, the root ostia, one for the sensory and one for the motor ramus, separated by the interradicular septum of dura mater. At this level the layer of pia arachnoid is much thickened and is adherent to the roots and the dural lining, thus sealing off the cerebrospinal fluid from passing further along the radicular nerve. Beyond the ostia the roots are carried in separate arachnoidal and dural compartments known as the root sleeves, which extend out for a short distance, and then the arachnoid ceases and the roots are invested only with the dural sheath. The two roots fuse just lateral to the posterior root ganglion and subsequently separate into posterior and anterior primary rami. It will be seen that the anatomical distinction between the root pouch and root sleeve is a real one, and together they

comprise what was formerly and loosely talked of as the root sheath. The segregation of the rami into separate dural compartments assumes importance in the operative treatment of brachial neuritis if the condition of root sleeve fibrosis is present.

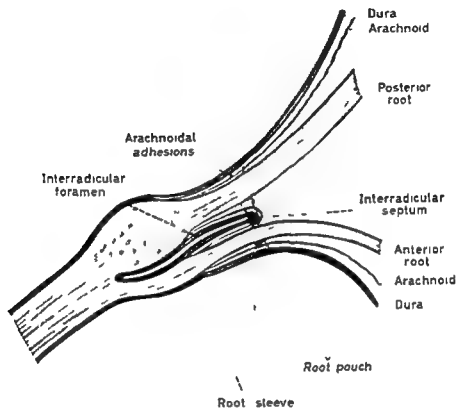


FIG. 120 —Diagrammatic drawing to show the anatomy of the root sleeve and root pouch (After Frykholm)

Root sleeve fibrosis

It has long been recognized that spondylotic protrusions lead to fibrotic changes in the dural sheaths surrounding the emergent roots often causing adhesion of the root to the surface of the protrusion but Frykholm has found that these changes which he terms root sleeve fibrosis are more specific and possess characteristic appearances which he has observed repeatedly at operation—the main features of which are as follows: (1) opacity of the dura of the root sleeve and root pouch; (2) narrowing and abolition of the root pouch; (3) thickening of the dura surrounding the root ostia and of the interradicular septum producing a constriction ring; and (4) fibrosis of the arachnoid in the vicinity of the root ostia exacerbating the effects of (3).

The presence of root sleeve fibrosis is of importance in treatment because although in a number of patients simple decompression of the nerve may be sufficient to relieve symptoms in those cases with well marked root sleeve fibrosis decompression is not enough and division of the dural compartments of the sensory and the motor roots may be required for complete relief.

SPINAL CORD INVOLVEMENT—CERVICAL MYELOPATHY

The term cervical myelopathy is an all embracing title used to describe the neurological results of damage to the cervical spinal cord, often in conjunction with the nerve roots by annular protrusions

The consequent clinical picture is pleomorphic the result of a number of inter-playing factors. As already mentioned the spinal cord may be involved alone or in company with one or more nerve roots and the symptoms referable to the nerve roots may precede or accompany or follow the appearance of cord symptoms. The course is variable in nearly a quarter of the cases the disease follows steadily and inexorably its downward path, whereas in others the deterioration tends to be slow, but episodic new symptoms and signs appearing at each episode and persisting. In a small number of patients the static interval between episodes may last for months or even years but any real improvement, when measured objectively is rare. It is this episodic nature of the condition, with intervals of spontaneous arrest that renders the critical appraisal of the results of treatment so very difficult.

Neurological signs

The dominating feature of the condition and usually the first to appear, is weakness and spasticity of one or both legs with the associated signs of exaggerated reflexes and extensor plantar responses. Sensory symptoms in the legs and trunk are less common and comprise paraesthesiae sometimes very uncomfortable and described as painful and reduction of pain and temperature sensation, often of a patchy distribution and rarely with a sharp upper level such as is seen with compressions by a cervical tumour. Light touch is not so commonly affected and posterior column sensation is usually preserved. The upper limbs present a much more diverse picture dependent upon the relative balance of spinal cord versus nerve root involvement. Thus there may be weakness and wasting which is limited to a single root distribution or confined to the small muscles of the hand on one or both sides or it may be generalized throughout the arms occasionally with fasciculation of the affected muscles. With the weakness and wasting there may be lack of tone and the reflexes may be reduced or absent but on the other hand the limbs may be spastic with very brisk reflexes. It is well recognized that the signs of nervous involvement particularly those related to the pyramidal tracts, may extend for a few segments higher than the actual site of compression.

The sensory signs in the arm vary considerably. In some patients there may be no changes whatsoever. In others diminished sensitivity to pin prick and light touch may be observed over dermatome areas of one or more adjacent nerve roots. It is most frequent on the inner border of the hand and forearm. In others the loss may be confined to the hand or forearm of a glove distribution probably from interference with the tracts in the spinal cord. Very occasionally there is dissociated anaesthesia over the upper limbs and the trunk resembling that of syringomyelia.

Sphincter disturbances occur in about one third of the patients are not severe and consist usually of hesitancy and urgency of micturition rarely incontinence. Disturbance of bowel function is exceedingly uncommon.

Factors concerned in producing the neurological picture

That cervical spondylosis should give a diverse neurological picture is understandable when consideration is given to the variable factors that may be concerned

First the level of the disc protrusion is important. Thus two, three or even the whole six cervical discs may be the site of protrusion causing multiple compressions of the cord with or without the related nerve roots. Again the situation of the osteophytic bar has an important influence depending on whether at or above the cervical cord enlargement in determining to what extent and in what manner the arms are involved. This is only one part of the explanation of the diverse picture and other factors have to be sought to clarify some of its puzzling features. For instance the neurological damage often extends to a level in the spinal cord higher than the disc protrusion. And another feature is that the pyramidal tracts are invariably, and usually predominantly, involved often without change in the spinothalamic tracts although these have a much closer relationship to the osteophytic bar than do the pyramidal tracts. Another aspect germane to this problem is that evidence of a substantial spinal block is rarely found, either on lumbar puncture or on myelography. A related observation is that usually little improvement in neurological signs may be expected from surgical treatment, the best that can be hoped for is arrest of deterioration. With true compression of the spinal cord by a benign tumour the exact converse holds true: there is usually a complete or almost complete block and the degree of neurological recovery even when there is an almost complete transection, is remarkable good.

Pathological cause

All these features taken together would suggest that simple compression of the spinal cord and its fibres is not the mechanism wholly responsible for the neurological signs in cervical myelopathy and we have to look elsewhere for the pathological cause for the spinal cord damage. A number of views have been put forward. The first has a purely mechanical basis and has been advanced by Kahn (1947). It concerns the tethering effect of the dentate ligaments which prevents backward displacement of the cord by the osteophytic bar and produces an area of stress in the fibres lying beneath the attachment of the ligamentum denticulatum to the cord in which site lie the pyramidal tracts. Changes in the vascular supply have been investigated by Mair and Druckman (1953) who showed in four necropsy specimens that demyelination was greatest in the territory of supply of the anterior spinal artery and its branches. Certainly both anatomically and pathologically the symptoms and signs could well be explained by ischaemia in this territory. It has been suggested that the damage to this vessel can be produced by repeated frictional injuries to its wall as the cord is pulled over the surface of the protrusion by the flexion and extension movements of the cervical spine during normal use. In addition the oscillation of the spinal cord that takes place with arterial and respiratory pulsation may play a part in causing frictional damage. Although it is unlikely that true compression plays much part in cervical myelopathy nevertheless intermittent compression may come about in two different ways. Taylor (1953) has attributed it to the ligamenta flava. When the head is extended the ligaments bulge forwards into the spinal canal as corrugations which can be shown up by myelography. The spinal cord displaced forwards by the ligament can be squeezed against a spur in front so that the nerve fibres become compressed and it is suggested that these repeated intermittent compressions are responsible for the signs. In some cases a spondylotic disc may be abnormally mobile so that the vertebra above can move forwards a considerable distance when the neck is

flexed, thus narrowing the canal and causing intermittent compression of the cord between the lamina of the vertebra above and the body of the one below.

A reasonable interpretation of present views is that the ultimate cause of cord damage is probably interference with the blood flow in the anterior spinal artery and its radicals, and that this may be brought about either by intermittent compression by the ligamentum flavum or by frictional injuries of the vessels during neck movement.

Differential diagnosis

The varied clinical picture of cervical myelopathy simulates other diseases of the central nervous system prevalent in middle age, and of these the most important are disseminated sclerosis, subacute combined degeneration of the cord, syringomyelia, extramedullary and intramedullary tumours of the spinal cord, and motor neurone disease. Another cause of confusion may be the coincidental finding of radiographic changes of cervical spondylosis in association with one of these diseases, and in this respect it is important to emphasize that the presence of radiographic changes of spondylosis does not necessarily imply that they are responsible for the neurological symptoms. The diagnosis of cervical myelopathy may present many problems, and in arriving at a correct interpretation of the physical signs the skill even of an experienced neurologist may be severely taxed. It is imperative to seek such a person's aid in confirming the diagnosis certainly before treatment by operation is contemplated.

Investigations

Plain radiographs

These should be taken in the antero-posterior and lateral projections, with oblique views from each side to show the degree of encroachment on the intervertebral foramina. In addition films of the spine in flexion and extension are needed to show any abnormal mobility of the discs. There may be revealed congenital anomalies such as fusion of one or more vertebral bodies, often with the spondylotic change lying above or below the site of the abnormality. The typical osteophytic bar with narrowing of the related disc will be seen at one or more levels but the actual situation of the spur, its relationship to the theca, and its height, can be reliably estimated only by myelography. The degree of narrowing of the disc is not necessarily an index of the extent of the spur formation (Fig. 121) and an extreme thinning of the disc may not be associated with any protrusion. Subluxation of one vertebra on another may be observed particularly on the flexion and extension films. There may be a disturbance of the vertical alignment of the spine. Usually the normal cervical curve tends to be exaggerated but sometimes it is obliterated or reversed so that the spine may be straight or even kyphotic and it can then be inferred that the cord is stretched forward across the ridge and that the slightest increase in the height of the protrusion will produce a disproportionate increase in tension on and indentation of the spinal cord and its vascular supply.

Spinal puncture

As an ancillary investigation in the diagnosis lumbar puncture has little positive value. Its main use is in revealing a complete manometric block and the high protein content in the cerebrospinal fluid due to the presence of an unsuspected tumour.

Usually in cervical spondylosis the manometrics are quite free but occasionally a partial or complete block may be shown up often only when the head and neck are extended. The protein content is quite normal in more than half the cases and in about a quarter there may be a rise in protein from 40 milligrams per cent up to on rare occasions 140 milligrams per cent.



FIG. 121.—Radiographs to show congenital partial fusion of the fourth and fifth cervical vertebrae with mild spondylotic narrowing of the C 3/4 disc. The myelogram however reveals a considerable intra spinal protusion which was confirmed at operation.

Myelography

This is a necessary investigation for final confirmation of the diagnosis of cervical myelopathy due to spondylosis and is an essential preliminary to operation either for myelopathy or for brachial neuritis from a lateral protrusion. It is important during the process of myelography to have several of the disc spaces covered at the same time so that a comparison of the extent of indentation may be made. To this end 5 or even 6 millilitres of Myodil are required introduced either by cisternal or lumbar injection. With intraforaminal and dorso lateral protrusions causing nerve root compression there is characteristically obliteration of the root pouch the filling defect having a curved medial border. These appearances may be found alone or as part of a ridge like filling defect which extends medially for a variable distance and represents an osteophytic bar.

With the central protrusions causing cervical myelopathy, a transverse filling defect of varying width will be seen and with really large osteophytic bars a

tent like lifting of the anterior thecal wall will occur, producing an elongated oval defect resembling that caused by an intramedullary cord tumour, but in the lateral views the extradural origin of the indentation will clarify the diagnosis.

Although myelography usually confirms the changes seen in the plain radiographs occasionally the findings are at variance. A common discrepancy arises from the fact that osteophytes seen in the plain radiograph and seemingly projecting into the canal may in fact be laterally situated and not in contact with the spinal cord or theca at all. Again an apparently small spur seen on the plain films may be shown on myelography to be much larger because of a cap of fibrocartilage. Myelography is of great value when several discs are the site of spur formation because it will determine which is the most prominent and which causes the greatest indentation.

Measurement from the anterior margin of the Myodil column to the posterior border of the body of the vertebra will indicate the height of the osteophytic bar but this by itself may not be so important as the amount of narrowing of the spinal canal it produces. It is this that renders possible the intermittent compression of the spinal cord by the corrugations of the ligamenta flava. This degree of narrowing of the canal is found by measuring from the anterior border of the column of Myodil to the posterior wall of the canal which is shown by the line of cortical bone where the spinous process joins the lamina. The average antero-posterior diameter of the spinal canal in the region of the lower three cervical vertebrae is 17 millimetres but this may vary by several millimetres (Wolf, Khilnani and Malis 1956). With a large spondylotic bar the canal may be narrowed to 10 millimetres or less. In a case with such a degree of narrowing it will be realized that as the antero-posterior diameter of the spinal cord at the lower cervical level averages 8 millimetres, the bulging of the ligamentum flavum must inevitably compress the cord albeit intermittently. In a personal series of cases with moderate to severe myelopathy the average measurement of the canal was 9.5 millimetres whereas the similar measurement at normal discs above or below was on average 6.5 millimetres greater. On the other hand as Wolf has suggested it seems unlikely that compression of the cord even intermittently can occur in a spinal canal whose antero-posterior diameter is greater than 12 or 13 millimetres as measured in this manner allowing even for the largest cord and the largest bulge of the ligamentum flavum.

TREATMENT

Dorso-lateral and foraminal protrusions

Conservative treatment

Brachialgia or brachial neuritis in a mild form is a very common complaint in middle age and in a large number of cases the patient treats himself by resting the arm or is given some gentle physiotherapy, with entirely satisfactory results. In the more acute case particularly if the pain persists for more than a few weeks and begins to wear down the patient's morale he is best treated in hospital with strict recumbency and immobilization of the neck between sand bags or in a felt or sponge rubber collar. With the knowledge of the underlying pathological changes responsible for the symptoms in these patients—that is to say a hard osteo-cartilaginous protrusion situated dorso-laterally or in the intervertebral foramen

and which has probably been present for months or years—it might be expected that traction on the neck would have no place in treatment. However in a proportion of cases halter traction with a moderate weight (4–8 pounds) is of help in the acute phase and tends to shorten the attack. In some cases it is of no benefit and in a few traction will tend to exacerbate the pain and will have to be discontinued.

Once the pain has subsided—and most attacks run their main course in 3–4 weeks—the patient can be got up in stages wearing a plastic moulded collar which can gradually be dispensed with once all his pain has disappeared. The severity of the pain associated with the lateral annular protrusions in cervical spondylosis tends not to be so severe as with the less common soft nuclear prolapse and it is possible to continue treatment for a much longer time. This fact coupled with the knowledge of the degenerative nature of the vertebral and disc changes means that surgery will be postponed until a really prolonged course of conservative treatment has failed.

Indications for surgery

The indications for surgery therefore are (1) resistance to adequate conservative treatment, and this will rarely be less than 3 months, except with unusually severe symptoms. (2) secondary attacks of pain within a short space of time, this indication will vary with the severity of the attacks and the patient's capacity to withstand pain, but usually three attacks within 12 to 18 months make the patient anxious for some more radical form of treatment, (3) the rare case of a disc protrusion that produces severe motor weakness of a segmental group of muscles often without much sensory change. This paralysis constitutes a very great disability in the upper limb.

Treatment by operation

As the protrusion in these cases is usually a hard osteo cartilaginous one it is not as a rule possible to remove it and, in fact the retraction of the nerve root necessary to give surgical access to the anteriorly situated osteophyte may add considerably to the damage to the nerve. Removal of the bone surmounting the root is the procedure usually adopted so that the nerve is no longer compressed. In addition in some cases with pronounced root sleeve fibrosis division of the dural sheath may be necessary (see below). The precise technical approach will vary according to whether a dorso lateral or an intraforaminal protrusion is present. In the former case as shown in Fig. 122 a partial hemilaminectomy should remove all the overlying bone that is capable of compressing and distorting the nerve. In the case of an intraforaminal protrusion this procedure will not give adequate decompression and the bone extending out into the intervertebral foramen has to be removed by the operation of partial facetectomy and the nerve uncovered actually in the canal. This procedure will have the same effect on the nerve whether it be compressed by an actual protrusion from the disc or by osteoarthritic lipping of Luschka's joints.

The surgical approach is the same for both methods—a separation of the muscles from the spine and laminae on the side to be explored. The laminae are then best identified by counting down from the prominent and unmistakable spine of the axis and if the operation is one of decompression by laminectomy the relevant half lamina is removed. The ligamentum flavum is excised and the root gently exposed.

TREATMENT

and retracted to reveal the disc protrusion. If by chance there is a soft cartilaginous cap present that lends itself to removal so much the better. However if a hard spur is present and this is usually the case it is best left undisturbed.

The performance of hemifacetotomy is best achieved with a 1.5 centimetre burr which is placed laterally on the lamina and the opening it produces exposes the medial aspect of the zygapophysial joint and finally the thinned roof of the foramen. This bone is flaked off and the opening into the spinal canal enlarged medially with rongeurs until the whole course of the nerve from the theca to the posterior root ganglion is exposed. The spur will be seen after retracting the root. This with removal of a cartilaginous cap if present is usually all that need be done but if there is

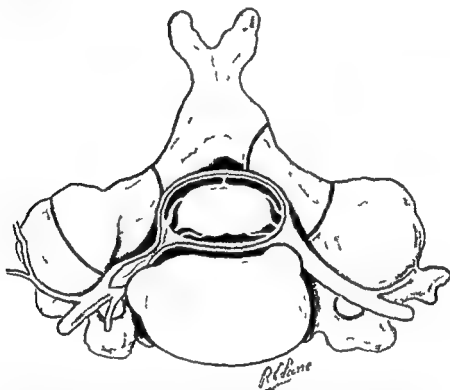


FIG 122 —Diagrammatic drawing which demonstrates the approach and decompressive effect afforded by hemilaminectomy (on the right) and hemifacetotomy (on the left) for the dorso lateral and intraforaminal protrusion respectively

evidence of root sleeve fibrosis then decompression of the two rami constituting the root will be needed. This will consist in making an incision in the long axis of the nerve from the root pouch laterally for 8 to 10 millimetres making sure the constricting ring at the root ostium is divided. This will expose the sensory ramus which is now freed from the adhesions binding it to the dural sleeve and retracted to disclose the anterior wall of the sleeve which is again incised thus opening into the interradicular foramen and revealing the posterior wall of the motor root sleeve which is incised in similar fashion the motor root then being freed from its adhesions. In this way both rami will be completely decompressed. The arachnoidal pouch will of course be opened in this procedure and it has to be borne in mind that if sepsis should occur the risks of operation are rendered more serious.

Results—The results of operation for lateral protrusion causing brachial neuralgia are very difficult to assess because few series have been published and even fewer in which a distinction is made between those cases with a soft nuclear

herniation and those with the annular protrusion with which we are concerned. The most carefully analysed results are those of Frykholm (1951) in his original communication. Thus of his 25 patients with annular protrusion who were kept under observation for not less than two years 15 showed considerable improvement, 5 showed moderate but persistent improvement and 5 were either unchanged or worse. It would seem that this 60 per cent chance of a good result is reasonable when dealing with such a chronic pathological condition as spondylosis, although not as good as the results obtained with the soft nuclear herniation. The present author with a smaller series of cases treated by laminectomy or partial facetectomy mainly without division of the root sleeves had similar results, with worthwhile improvement in about 65 per cent of cases.

Radiotherapy

Although radiotherapy of the cervical spine is inferior to surgery for treating the younger patients it is a form of therapy worth considering for the very old patients because the dosage required is not very great and should not produce much constitutional disturbance, in a considerable number of cases it eases the pain although often only temporarily. Lundar (1951) has reported upon 625 patients treated by radiotherapy of whom 70 per cent were stated to have benefited considerably, but in nearly 50 per cent symptoms returned.

Cervical myelopathy

Usually the progress of the disease is slow deterioration being observed over periods measured in months or even years so that treatment may have to be prolonged and equally it may take a long time to assess its value in arresting progress. Occasionally deterioration is rapid from the beginning or having started slowly the symptoms have increased rapidly and operation may become a matter of some urgency in attempting to prevent the more severe consequences.

Conservative treatment

In most cases however the basis of treatment initially will be strict bed rest and immobilization of the neck. This can be achieved by fixation of the head between sand bags or by halter traction to the head which acts mainly by preventing flexion movements rather than by any distracting effect. Physiotherapy to the limbs is continued during this phase. The response may be quite gratifying. Within a short time power may improve and sensory change becomes less evident but usually falling far short of normal. In other patients there may be only an arrest of the progress of the crippling condition and again in others no observable change occurs. Bed rest is continued in the absence of any deterioration, for 3-6 weeks and the patient then gets about wearing a moulded plastic collar the main purpose of which is to restrict rather than prevent cervical movements. It is hoped by these means to preserve the improvement gained by recumbency. This collar is worn for a time related to the response obtained. Usually at least 3 months is advised in those who have shown a good response in others with no more than a halting of symptoms it will have to be worn for many months and then discarded gradually with careful watch for any neurological deterioration and in a few cases the collar will become a permanent part of the patient's dress. The value of this initial conservative treatment is not yet accurately known despite the large number of patients who must by now have been treated in this fashion and no large series with

prolonged observation has yet been published Brain Northfield and Wilkinson (1952) reported some improvement in 11 of 14 patients treated conservatively In 16 patients treated solely by neck immobilization 8 showed improvement and in 2 of them it was quite striking (Clarke and Robinson 1956)

Treatment by operation

The main indication for operation in this condition is progressive deterioration in spite of adequate conservative treatment Occasionally for a patient in whom the symptoms are becoming rapidly crippling surgery may have to be considered without delaying for the several weeks necessary to assess the outcome of conservative methods

There are two main principles of operation directed primarily at aiding the myelopathy The first and the most commonly employed is laminectomy with removal of the ligamenta flava and thus decompression of the posterior aspect of the theca to which procedure there may be added the opening of the dura mater with division of the dentate ligaments above and below the disc level This type of operation will permit the spinal cord to move backwards away from the osteophytic bar and thus escape to a varying extent the frictional and compression injuries caused by neck movements The other method of treatment is a direct exposure of the osteophytic bar and its removal from in front of the theca and spinal cord

Laminectomy—In order to obtain a satisfactory decompression the laminae above and below the related disc with their attached ligamenta flava require to be removed and this holds also for multiple disc protrusions Thus for three disc protrusions at adjacent levels four laminae will need to be excised In addition it is sometimes the practice to open the theca and divide the ligamentum denticulatum on each side as far as the exposure will allow in the belief that harmful traction on the cord may be prevented and also that a greater displacement of the cord backward will be permitted This procedure will carry of course a slightly greater risk in the event of infection, and also in some patients with severe cord damage the inherent manipulations of the spinal cord may cause profound neurological deterioration

The results of this type of operation have proved difficult to assess In the first instance cervical myelopathy is of comparatively recent recognition and so there has been no prolonged follow up of surgical cases Secondly there are a number of imponderables such as the tendency for spontaneous arrest the fact that the bed rest entailed by surgery may itself cause some improvement and finally the difficulty in establishing criteria of partial improvement

The largest personal series is that of Northfield (1955) whose results in 39 cases of laminectomy with dentate ligament section are as follows

Deaths	1
Slowly worsened	8
Stationary	8
Slight improvement	9
Considerable improvement	13

There have been reports of other and smaller series a selection of which has been set down in the Table following These reports were chosen as incorporating enough information about the cases and the neurological and operative findings to make a reasonable assessment possible

TABLE
SHOWING THE IMPROVEMENT (IF ANY) FOLLOWING LAMINECTOMY

Author	Improvement			Unchanged or worse	Total
	Considerable	Moderate	Slight		
Northfield (1955)	13	—	9	16	38
Walsh and Mackenzie (1956)	—	13	4	11	28
Arnold (1955)	2	2	1	3	8
Epstein and Davidoff (1951)	1	1	—	2	4
Seegerberg (1956)	3	4	—	—	7
Totals	19	20	14	32	85

It will be seen that there is considerable variation in the results of different authors but roughly 39 or a little under half, of the patients derived worthwhile improvement from surgery. In no case was a return to normal function obtained.

Excision of the protrusion—The more radical approach, removal of the osteophytic spur, has been advocated by Allen (1952). This procedure consists in a formal laminectomy with exposure of the protrusion and the related nerve roots. The posterior longitudinal ligament is then divided in cruciate fashion above and below the nerve root, this being gently retracted to expose the bony surface of the spur. With a special curved chisel the ridge is then removed piece meal through a natural plane of cleavage which can be distinguished between the base of the osteophyte and the posterior surface of the body of the vertebra. Working from each side and from above and below the nerve root it is possible to remove the spur in its entirety.

Despite the risks of working in front of a seriously damaged spinal cord with impaired blood supply Allen has been able to carry out the procedure without evidence of further harm. A detailed assessment of the results, particularly long term, of this type of operation have not as yet been published.

Graft operations—Besides these two main surgical procedures other methods have been employed to a lesser extent. Occasionally in cervical spondylosis the flexion extension films reveal excessive mobility at one particular disc level so that the upper part of the cervical column moves forward on the lower part to compress the cord mechanically. This subluxation can be treated by fixing the vertebrae above to those below by posterior bone grafts, possibly reinforced by wiring of the spinous processes. Posterior grafting of vertebrae as mobile as the cervical often fails, and so an anterior route has been devised, the vertebral bodies themselves being grafted. Through an oblique incision with retraction of the great vessels the graft is inserted into a channel cut into the vertebral bodies and the intervertebral discs. Another anterior method to promote intercorporeal fusion has been reported by Deraymaker and Gehuchten (1956) in 12 patients, 4 with brachialgia and 8 with myelopathy, of whom 9 were stated to have benefited. These are all major procedures which must still be regarded as somewhat experimental and which have been performed only in small numbers, few details and no prolonged follow up of the cases are available.

Conclusions—The precise role of surgery in the treatment of cervical myelopathy has yet to be established not only in relation to the type of operation but also to the selection of patients most likely to benefit. As a rough guide experience suggests that surgery stands its best chance of success in patients under the age of 50 years who have a comparatively short history of deterioration and in whom the spondylosis is confined to one disc level and has produced severe narrowing of the spinal canal. The operation most favoured at the present time is decompressive laminectomy associated perhaps with ligamentum denticulatum section. Just under half the patients selected for this procedure will show worthwhile benefit and without great risk. But it is not yet known what the long term results of such surgery will be or whether the weakening of the spine that may follow laminectomy will promote spondylosis at other levels. Surgery may prove to have, at most only a temporary effect. The future really lies in establishing the causes of the degenerative changes in the intervertebral discs leading to cervical spondylosis and preventing them, but this lies a long way ahead and the more immediate objective now is early diagnosis and the development of surgical techniques for removing the osteophytic bar itself from in front of the theca without undue risk.

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CHAPTER 10

TUMOURS OF THE VERTEBRAL COLUMN

H A SISSONS

INTRODUCTION

ALTHOUGH accurate information has not been published on the relative frequency of the different types of tumour known to involve the bony tissue of the vertebral column it is clear from most general accounts of bone tumours (Coley 1949, Geschickter and Copeland 1949, Lichtenstein, 1952) that this region contrasts in many respects with other parts of the skeleton and that it poses certain special problems concerning the recognition of neoplastic lesions.

With a few types of primary bone tumours including angioma, aneurysmal bone cyst and benign osteoblastoma, an important proportion of all recorded cases occurs in the spine and one primary bone tumour—chordoma—occurs only in the axial skeleton. Most types of primary bone tumour however, including benign chondroblastoma, chondromyxoid fibroma, giant cell tumour, osteosarcoma, chondrosarcoma and fibrosarcoma, are less often encountered amongst series of spinal tumours than their frequency in other parts of the skeleton might suggest. Metastatic tumours however often involve the spine and account for the great majority of cases in any representative series of vertebral tumours in adults. This should be remembered in the differential diagnosis of a case of suspected vertebral tumour.

In the absence then of dependable information about the relative frequency of the different varieties of bone tumour affecting the spine this chapter will outline the general features of each type of lesion, giving particular attention to entities which have been recognized in recent years. Emphasis is given to pathology, because of its importance in helping to define the various types of tumour with which we are dealing.

These are discussed under the following headings:

Secondary tumours: Metastatic carcinoma

Tumours of haemopoietic and reticuloendothelial tissue: Myelomatosis, leukaemia, lymphosarcoma and Hodgkin's disease

Primary tumours of bone tissue: Malignant—Osteosarcoma, chondrosarcoma, fibrosarcoma, giant-cell tumour and chordoma. Benign—Angioma, aneurysmal bone cyst and benign osteoblastoma.

Intraspinal tumours

SECONDARY TUMOURS

Metastatic carcinoma

The general features of secondary tumours of bone are admirably described by Willis (1934).

The skeleton is a frequent site for metastasis from many types of malignant epithelial tumour. In a series of 500 carcinoma cases studied at autopsy Willis (1934) showed that bone metastases occurred in 15–25 per cent of all cases; the

skeleton being involved only less frequently than the lungs and liver. A similar incidence (27 per cent) was found in a larger series of cases reported more recently by Abrams (1950). Some types of primary tumour however, metastasize to bone much more often, examples being carcinoma of the breast (about 50 per cent of fatal cases) and of the prostate gland (about 70 per cent of fatal cases). According to Abrams (1950) and to Meyer (1957) the primary tumours most often responsible for skeletal metastases are carcinomas of the breast, prostate gland and lung, followed by carcinomas of the colon, stomach and bladder, all of which are less rarely responsible for bone secondaries than is generally supposed, and some way down the list are carcinomas of the thyroid gland and kidney. The primary tumour responsible for skeletal metastases is often clinically latent. Carcinomas of the thyroid gland and the kidney have a special reputation in this connexion.

Metastatic bone tumours almost invariably involve the haemopoietic or red, bone marrow, which in the adult is normally present in the vertebral bodies, the pelvis, the calvarium, the ribs and sternum, and the proximal ends of the femur and humerus. The spine, in fact, is one of the most frequent sites for metastatic bone tumours, the lumbar vertebrae being rather more commonly involved than others.

The metastases are blood borne, and it is most likely that they reach the bone marrow as emboli in the systemic arterial system. The vertebral plexus of veins has been suggested as a pathway for tumour metastasis from the pelvic viscera—the prostate gland in particular—to the bodies of the lumbar vertebrae, but its role is unproven and would seem unlikely. While injection studies (Batson, 1940; Franks, 1953) established the existence of this plexus, it has not been shown that tumour emboli from the pelvic viscera would normally enter it, or that having entered it they would lodge in the marrow sinusoids instead of returning to the caval circulation. With metastasis to bones, as with metastasis to other organs, the observed distribution of tumours is to be explained by the generalized dissemination of tumour emboli and by their selective growth in certain territories, in this case haemopoietic bone marrow.

Spinal metastases are usually multiple, but solitary or initially solitary lesions are encountered and often simulate primary bone tumours. The metastases may be circumscribed or diffuse (Figs 123, 124). A circumscribed deposit, if small, is a spherical mass involving part of a vertebral body. It grows to occupy the entire vertebral body and may occasionally form a soft tissue mass involving adjacent structures, although the intervertebral disc tissue usually acts as a barrier to its growth. Most secondary deposits are associated with destruction of the invaded bone and are consequently termed *osteolytic*. In the spine such bone destruction commonly results in vertebral collapse and spinal cord compression. Some tumours, particularly those of prostatic origin, stimulate new formation of bone (Figs 124, 125); they produce dense radio opaque lesions and are termed *osteosclerotic*. Bone formation in metastatic tumours is due to osteoblastic activity (Fig 125). Bone destruction is usually the result of osteoclastic activity, although it has been suggested (Milch and Changus, 1956) that it may sometimes be caused by the tumour cells themselves. Although the terms osteolytic and osteosclerotic are convenient, they denote differences of degree, bone destruction and bone formation both being present in most lesions. Bone metastases

CHAPTER 10

TUMOURS OF THE VERTEBRAL COLUMN

H A SISSONS

INTRODUCTION

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neuroblastoma is one of the more common varieties of malignant skeletal tumour encountered. Primary malignant bone tumours particularly osteosarcomas may themselves metastasize to bone—although they do so infrequently—and it is sometimes difficult (Sissons 1958) to distinguish between such metastases and multifocal primary tumours.

TUMOURS OF HAEMOPOIETIC AND RETICULO ENDOTHELIAL TISSUE

Myelomatosis

After metastatic carcinoma this is one of the commonest neoplastic conditions involving the spine. In common with other haemopoietic neoplasms it shows diffuse or multifocal origin. Evidence of generalized bone marrow involvement is usually present from the beginning and lesions may also develop in other organs, particularly the spleen (Churg and Gordon 1950) without the ordinary



(a)



(b)

Fig 126—Myelomatosis. (a) Radiograph showing collapsed vertebra. (b) Longitudinally divided thoracolumbar spine from the same case. The posterior extension of the tumour has produced cord compression and paraplegia. The other vertebrae shown appear normal but there were disseminated skeletal lesions elsewhere.

manifestations of blood borne metastasis. Occasionally a tumour with the histological structure of myeloma appears to be solitary, but experience makes it clear that other lesions either exist or can be expected to develop (Christopherson and Miller 1950 and Raven and Willis 1949 who also discuss the few authenticated cases of solitary myeloma).

Myelomatosis usually occurs in individuals above 50 years of age, men being more often affected than women. It is often associated with abnormalities of plasma proteins in the form of increased serum globulin with certain abnormal β or γ globulin components as shown by electrophoretic analysis and with continuous



FIG 123—Isolated osteolytic metastasis from carcinoma of kidney. Vertebral collapse has caused compression of the spinal cord. Invasions of soft tissues has occurred and the tumour has extended from one vertebral body to that immediately above. (Specimen from museum of Royal College of Surgeons of England)

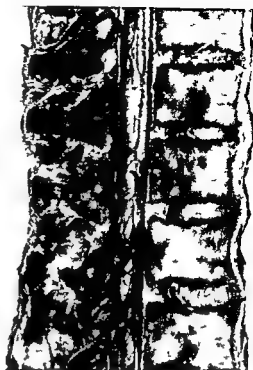


FIG 124—Diffuse osteoclerotic metastases from prostatic carcinoma. The external contour of each vertebra is normal but all are diffusely infiltrated by tumour tissue and converted into abnormally dense bone. (Specimen from museum of London Hospital Medical College)



FIG 125—Osteosclerotic metastasis from carcinoma of pancreas. Scattered glandular structure of the tumour are separated by a close network of bony tissue. A few pre-existing trabeculae of lamellar bone are present but the remainder is newly formed non-lamellar bone produced by osteoblastic activity (x48)

moreover are not always associated with alteration of bone structure. As shown by Shackman and Harrison (1948) gross tumour deposits may be present without any demonstrable radiographic change.

Non epithelial tumours such as soft tissue sarcomas and lymphosarcomas are occasionally responsible for skeletal metastases. In children metastatic

neuroblastoma is one of the more common varieties of malignant skeletal tumour encountered. Primary malignant bone tumours particularly osteosarcomas, may themselves metastasize to bone—although they do so infrequently—and it is sometimes difficult (Sissons 1958) to distinguish between such metastases and multifocal primary tumours.

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Myelomatosis usually occurs in individuals above 50 years of age, men being more often affected than women. It is often associated with abnormalities of plasma proteins in the form of increased serum globulin with certain abnormal β or γ globulin components as shown by electrophoretic analysis and with continuous

or intermittent proteinuria of a distinctive kind (Bence Jones proteinuria). These biochemical abnormalities are well reviewed by Rundles, Cooper and Willett (1951). It has been shown by Martin (1947) and by Miller and his colleagues (1952) that the myeloma tissue itself contains the same abnormal globulins that are present in the blood.

The usual skeletal lesions consist of disseminated areas of soft tumour tissue, producing focal bone destruction and having a sharply demarcated punched out appearance in radiographs. Spine, pelvis, ribs, sternum, skull and occasionally other bones are involved. In the spine, focal bone destruction often leads to



FIG 127—Myelomatosis. Longitudinally divided thoracic spine. Focal bone destruction is not present but there is diffuse replacement of bone marrow by myelomatous tissue. Considerable bone rarefaction is present and the bodies of several vertebrae show some collapse.

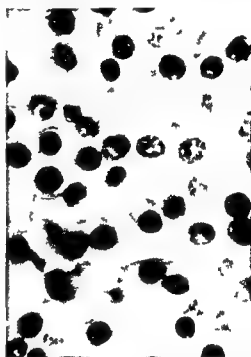


FIG 128—Myelomatosis. A high power view of part of a focal lesion illustrating the general resemblance of the myeloma cells to plasma cells (x 1110).

collapse of one or more vertebral bodies (Figs 126a, b). Such lesions are obvious radiologically but it is important to remember, as emphasized by Naylor and Chester Williams (1954), that the spinal abnormality may occasionally take the form of a less conspicuous diffuse bony rarefaction sometimes associated with vertebral collapse (Fig 127).

The tumour tissue is made up of cells whose exact nature is uncertain but which have a definite and characteristic morphology and show certain similarities to plasma cells (Fig 128). The myeloma cells are small rounded structures with clearly defined cytoplasm; their nuclei show a clumped arrangement of chromatin. It is unusual to see any reactive bone formation either in or at the margin of the lesions.

PRIMARY TUMOURS OF BONY TISSUE

Clinically, distinction from metastatic carcinoma is often possible through the recognition of the biochemical abnormalities mentioned above and the diagnosis can be confirmed by examination of sternum marrow or by biopsy of one of the bone lesions.

Paraplegia, from vertebral collapse with cord compression is an important complication of spinal lesions. Other complications of myelomatosis include anaemia, amyloidosis and renal failure. When present, amyloid deposits involve the myocardium, blood vessels, skeletal muscle and gastrointestinal tract (Snapper, Turner and Muscovitz, 1953) being comparable in their distribution with 'primary' amyloidosis and not with the 'secondary' amyloidosis that occurs in chronic suppurative conditions in which the liver, spleen and kidney are usually involved.

Treatment and prognosis

Radiotherapy is the accepted method of treatment in myelomatosis. Although it may have a dramatic though transient, effect on individual lesions, it has according to the series of cases studied by Garland and Kennedy (1948) only a slight effect in prolonging life, the condition usually proving fatal within a year or so of recognition. There has been considerable interest in recent years in the occasionally dramatic effects of chemotherapeutic agents in myelomatosis, particularly urethane (Rundles, Dillon and Dillon, 1950) and stilbamidine (Snapper, 1948).

Recent reviews of myelomatosis include that of Snapper, Turner and Muscovitz (1953) (clinical aspects) and those of Lichtenstein and Jaffe (1947) and Lumb (1952) (pathology).

Leukaemia, lymphosarcoma and Hodgkin's disease

Spinal lesions occasionally occur in leukaemia, lymphosarcoma or Hodgkin's disease, but they are rare. When encountered, the bone lesions are usually multiple, and are often clearly preceded by the more conspicuous and important soft tissue lesions. Occasionally a bony abnormality is the presenting lesion when it may be confused with other types of spinal tumour, particularly with metastatic carcinoma or myelomatosis.

PRIMARY TUMOURS OF BONE TISSUE

MALIGNANT

Sarcoma and giant-cell tumour

Those varieties of malignant bone tumours most frequently encountered in other parts of the skeleton (that is, osteosarcoma, chondrosarcoma, fibrosarcoma and giant cell tumour) occur very rarely in the spine. Even in Paget's disease—an important precursor of malignant bone tumours and a condition that itself commonly involves the spine—spinal tumours are rare (Lake, 1951).

Older accounts include a substantial number of giant cell tumours of bone among tumours of the spine. In Geschickter and Copeland's (1949) series of 58 cases of spinal tumour, there were 15 giant cell tumours, and Murphy (1935) collected 45 cases from the literature. While a few of these cases may have been true giant cell tumours, the more stringent criteria now adopted for the diagnosis of giant cell tumour (Jaffe, Lichtenstein and Portis, 1940) and the recognition that many cystic giant celled lesions of the spine are to be regarded as a separate entity, aneurysmal bone cyst (see page 202) make it clear that others were not. Caution must be exercised before a diagnosis of giant cell tumour can be regarded as established for a spinal lesion.

Chordoma

Like other primary malignant tumours of the vertebral column chordoma is a relatively rare lesion : because of its special regional interest, however, its features will be described in some detail

Origin

Chordoma is a distinctive type of malignant bone tumour occurring only in the axial skeleton and considered to arise from developmental remnants of axial notochordal tissue. Such remnants are occasionally encountered in normal individuals—particularly in the spheno occipital region where they take the form of small flattened masses of soft tissue attached by a narrow pedicle to the inner surface of the dura. Their presence was originally noted by Luschka (1856) and they were described in more detail by Virchow (1857). Virchow regarding them as of cartilaginous origin, and noting in them vacuolated or physaliphorous cells used the term *ecchondrosis physaliphora* for these structures. But Muller (1858) suggested that the anatomical situation of the nodules and their histological similarity to notochordal tissue indicated that they were derived from this structure and not from cartilage. The idea was endorsed by Ribbert (1894) and has since been generally accepted. Ribbert used the term *ecchordosis* to describe the notochordal remnants and later applied the name chordoma to tumours regarded as having comparable origin. The first of these frank tumours comparable with the *ecchordoses* had been described by Klebs (1864) and a considerable number have subsequently been reported. Early reports include that of an invasive intracranial spheno occipital chordoma by Fischer (1907) and of a sacrococcygeal case by Feldmann (1910). Useful reviews of the subject include those of Dahlin and MacCarty (1952) Stewart and Morin (1926) Utne and Pugh (1955) and Willis (1953).

Age and sex incidence

Despite their origin from developmental remnants chordomas usually occur in patients aged from 40 to 70 years. In one case however reported by Ellis (1935) the patient was aged 8 years. The lesion was in the lumbosacral region, but was unusual in that it was intradural and not intraosseous. In this respect it resembled a spheno occipital *ecchordosis* rather than the usual sacral chordoma. Males are more often affected than females.

Site

Most chordomas are situated in the sacral region and the next most frequent site is the spheno occipital region of the skull base but lesions of the intervening vertebrae also occur. Sacral chordomas usually present as pelvic tumours while those of the spheno occipital region produce the signs and symptoms of an intracranial tumour together with evidence of involvement of structures at the base of the skull. With cervical thoracic or lumbar tumours compression of the spinal cord or of the nerve roots occurs.

Clinical course

Chordomas are slowly growing tumours. They are locally malignant infiltrating adjacent structures and recurring after local excision and almost invariably causing death by local involvement of the central nervous system. Metastasis to the lungs or lymph nodes is infrequent but has been reported occasionally (Willis 1953).

Macroscopic appearances

By the time it has produced symptoms a chordoma frequently appears as a large tumour associated with considerable destruction of bone (Figs 129-130). It may extend beyond the confines of the spine as a bulky mass, and although its

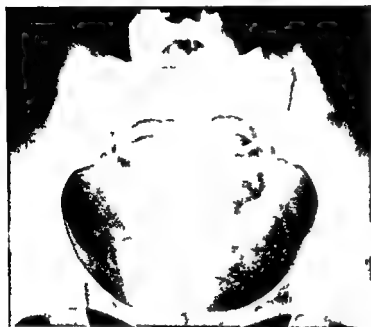


FIG 129—Chordoma of sacrum. Radiograph showing extensive bone destruction of the lower part of the sacrum.



FIG 130—Chordoma of sacrum. Longitudinally divided lumbosacral spine from another case: a bulky mass of lobulated tumour tissue replaces the sacrum and extends anteriorly and posteriorly.

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margin shows some degree of encapsulation there is often infiltration of adjacent muscle and other soft tissues. The tumour tissue itself is gelatinous and shows conspicuous lobulation (Figs 130 131*a*) areas of necrosis and haemorrhage are frequent



(a)



(b)



(c)



(d)

FIG 131 —Chordoma. These illustrations show different areas of the same tumour. (a) Low power view of histological section showing the characteristic lobulation of the tumour tissue ($\times 6$) (b) A field showing pleomorphic undifferentiated tumour tissue. Some cells have multiple nuclei ($\times 45$) (c) Part of a lobule of tumour tissue. The tumour cells are arranged in strands and clumps and there is abundant mucoid intercellular material ($\times 45$) (d) An area of tumour tissue showing conspicuous vacuolation ($\times 70$)

Microscopic appearances

It has already been noted that these tumours show a histological resemblance to notochordal tissue. They are characterized by large amounts of mucoid intercellular material in which are scattered strands and masses of tumour cells (Figs 131c, 132, 133). The tumour cells themselves are conspicuously vacuolated (Figs 131d, 133) giving the characteristic physaliphorous structures. These features are usually prominent but in occasional cases they are less evident. Smaller or larger areas of tumour tissue showing an undifferentiated pleomorphic appearance (Fig. 131b). Such absence of specific histological features occasionally leads to problems of diagnosis in small biopsy specimens, and in some cases the arrangement of the tumour cells in strands and cords may even suggest a diagnosis of



FIG. 132—Chordoma. Part of a lobule of tumour tissue showing strands of tumour cells separated by abundant mucoid intercellular material.



FIG. 133—Chordoma. A field where arrangement of tumour cells in cords might at first sight suggest an epithelial tumour. The tumour cells however show conspicuous vacuolation and abundant mucoid intercellular material is present.

metastatic carcinoma (Fig. 133). However, careful scrutiny of an adequate sample should reveal the definitive characteristics enumerated above and establish the diagnosis unequivocally. Occasionally the abundant mucoid intercellular material of a chordoma may show a superficial resemblance to the degenerated intercellular matrix of a chondrosarcoma; the cells of the latter however are rounded and lie in definite spaces in the matrix and they fail to show the conspicuous vacuolation of a chordoma.

Treatment

Chordomas are not usually regarded as radiosensitive though irradiation treatment is sometimes employed as a palliative procedure. Radical surgery is

at present the only hope for the ultimate survival of the patient but it can rarely be employed because of the intimate relationship of the tumours to the spinal cord and nerve roots

BENIGN

Angioma

Benign vascular tumours occur quite commonly in vertebral bodies but only rarely cause symptoms. Junghanns (1932) found them in about 10 per cent of a large series of necropsy cases in which the spine was carefully examined. Like other angiomas they are vascular malformations rather than progressively enlarging neoplasms. Despite this, however they are usually encountered in later life either on radiological examination or at necropsy. They occupy most



FIG 134—Angioma of spine. The bone marrow of the middle vertebra is replaced by cavernous vascular tissue—an incidental finding at necropsy (*Specimen from Department of Pathology University of Cambridge*)

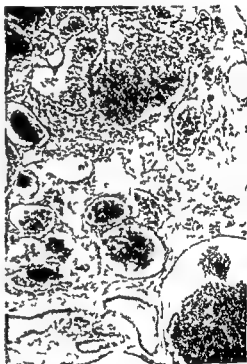


FIG 135—Angioma. The histological appearance of another lesion showing blood filled vascular channels of various sizes (x 43)

or all of a vertebral body and consist of thin walled cavernous blood vessels (Figs 134 135). Vertebral collapse and spinal cord compression have been described but are very rare. These lesions have been reviewed by Perman (1926) and by Macrycostas (1927)

Aneurysmal bone cyst

This is a distinctive although uncommon lesion which occurs in the spine and in other parts of the skeleton. It is a recently described entity and before its recognition by Jaffe and Lichtenstein (Jaffe 1950 Lichtenstein 1950) examples were probably regarded as haemorrhagic or cystic giant cell tumours

PRIMARY TUMOURS OF BONE TISSUE

There is no doubt, however, that the two conditions should be separated on the grounds both of structure and behaviour. Subsequent reports of aneurysmal bone cyst include those of Barnes (1956) Besse and his colleagues (1953) and Thompson (1954). Hadders and Oterdoom (1956) have suggested that aneurysmal bone cyst is a variety of angioma of bone.



(a)



(b)

FIG 136—Aneurysmal bone cyst. (a) Radiograph showing an expanding osteolytic lesion of the fifth lumbar vertebra and the adjacent part of the sacrum. (b) The lesion at operation. (B) courtesy of Mr Roland Barnes and Editor *J Bone Jt Surg*.)

FIG 137—Aneurysmal bone cyst. Histological appearance of tissue from the lesion shown in Fig 136. Large blood filled spaces are separated by thin septa of vascular fibrous tissue containing numerous giant cells (x35). (B) courtesy of Mr Roland Barnes.)



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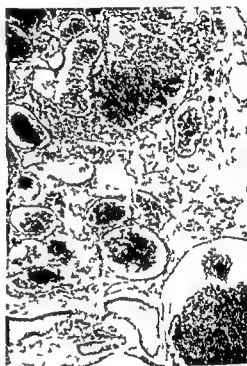


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Benign osteoblastoma

Terminology

Another newcomer to the ranks of benign tumours of bone is benign osteoblastoma. It is not uncommon among benign tumours of the spine but many names have been used to describe it and there is still some uncertainty in nomenclature. In 1952 the term 'osteogenic fibroma' was used by Lichtenstein to describe certain benign osteoblastic lesions. Additional cases were reported by Golding and Sissons (1954) and by Kirkpatrick and Murry (1955). Dahlin and Johnson (1954) reported a series of comparable cases using the term 'giant osteoid osteoma' to describe them. Recently Jaffe (1956) and Lichtenstein (1956) have each published general accounts of this tumour, and have independently suggested the term 'benign osteoblastoma' in order to distinguish it from osteoid osteoma and from various fibromas of bone.



FIG 139—Benign osteoblastoma. Radio-graph showing erosion of one lamina and pedicle of the first thoracic vertebra by irregularly calcified mass. Female aged 14 years. Golding and Sissons Case 1 1954. (B) courtesy of Editor *J. Bone Jt. Surg.*



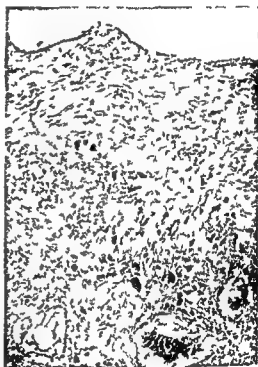
FIG 140—Benign osteoblastoma. Histological appearance of tissue from the lesion shown in Fig 139. It consists largely of uncalcified osteoblastic tissue but scattered darkly staining areas of calcification are present and some osteoclast giant cells are seen (x 115). (B) courtesy of Editor *J. Bone Jt. Surg.*

Most of the reported cases are in children or young adults, males and females being equally affected. Although encountered elsewhere the majority of tumours involve either the spine or the long bones of the hand or foot. In a vertebra the lesion is usually situated in the neural arch but is sometimes in the body. Clinical features are not distinctive: there is often pain and sometimes evidence of spinal cord compression. The radiological appearances (Fig 139) commonly take the form

Aneurysmal bone cyst occurs in older children adolescents and young adults. There is no predominance of either sex among the cases reported. Radiologically the lesion appears as a ballooned out distension of the periosteum, outlined by a paper thin subperiosteal bone shell (Jaffe, 1950) and it was this appearance that suggested the name aneurysmal bone cyst. Bone destruction may be conspicuous (Fig 136a) and the lesion may extend to involve more than one vertebra (Fig 136a). On surgical exploration the lesion appears as a blood filled cystic structure eroding bone and displacing the periosteum (Fig 136b) the central cavity is divided by ragged septa of reddish brown solid tissue. Histologically (Figs 137-138) the tenuous cyst lining and the septa consist of fibrous tissue



(a)



(b)

Fig 138 —(a) A field from a more solid region of another aneurysmal bone cyst. Newly formed bone and conspicuous blood vessels are present in the loose fibrous tissue. This type of vascular structure has given rise to the suggestion that aneurysmal bone cyst is a variety of angioma (x 40). (b) Another field from the same lesion. Osteoclast giant cells and newly formed bone are present in the fibrous tissue lining one of the large cystic cavities present (x 40). (By courtesy of Mr Roland Barnes.)

containing numerous vascular channels. Giant cells and haemosiderin pigment are often conspicuous in the fibrous tissue which also contains areas of scattered bone formation.

The rapid development of these lesions sometimes suggests that they are malignant but although they occasionally reach a large size they are entirely benign and excision or curettage is likely to produce complete cure.

DIAGNOSIS OF VERTEBRAL TUMOURS

malignancy is always lacking. In their behaviour, too, these lesions are always benign, despite the gravity of the symptoms they can produce by compression of the spinal cord or nerve roots. Treatment involves local resection of the lesion.

In the past these lesions have sometimes been wrongly identified on histological examination as giant cell tumours. With relatively few giant cells they have been called osteomas or even mistaken for osteosarcomas.

Other benign tumours

Chondromas and cartilage capped exostoses of vertebrae have occasionally been described involving vertebrae (Coley 1949, Geschickter and Copeland 1949). An exostosis may be a solitary lesion or part of hereditary multiple exostoses. Fibromas and lipomas are encountered. Osteoid osteomas of the spine, distinct from benign osteoblastomas, also occur (Jaffe 1945).

INTRASPINAL TUMOURS

It is necessary to distinguish between vertebral tumours proper and intraspinal soft tissue tumours. The symptoms of brackache and root pain, with evidence of spinal block, occur in each group, and clinical and even radiological differentiation between them is not always possible. It may not be until surgical exploration that the nature and site of the tumour is determined.

The series of intraspinal tumours reported by Elsberg (1941) and by Bloom, Ellis and Jennett (1955) indicate that benign nerve sheath tumours and meningiomas are much commoner than tumours of the spinal cord itself, which include metastatic carcinoma, glial and ependymal tumours, and occasional connective tissue tumours of uncertain origin.

DIAGNOSIS OF VERTEBRAL TUMOURS

As with bone tumours generally, radiological examination and biopsy examination are the two important aids to the diagnosis of lesions of the spine when a clinical diagnosis of tumour is under consideration.

Biochemical evidence is important in the recognition of myelomatosis (abnormal serum proteins, Bence Jones proteinuria), metastatic prostatic carcinoma (high serum acid phosphatase) and occasional osteoblastic lesions (high serum alkaline phosphatase). It may also direct attention to some generalized disease such as hyperparathyroidism when this is responsible for an apparently isolated bone lesion. Haematological examination and sternal marrow puncture sometimes provide important diagnostic evidence of neoplasms of bone marrow.

With circumscribed and presumably benign spinal tumours, surgical excision is usually the treatment of choice, and this procedure itself provides the entire lesion for histological diagnosis. When however there is clinical uncertainty about the possibility of malignancy, and when some other treatment such as radiotherapy is contemplated, elective biopsy has to be undertaken in order to establish the nature of the lesion. Open surgical biopsy, the most desirable procedure for the diagnosis of bone tumours in many other situations, is complicated by the inaccessibility of the deeply seated vertebrae; methods of aspiration biopsy

of an ill defined area of expansion of the involved bone irregularly distributed calcification sometimes being present. The diffuse bony sclerosis, so often seen in relationship to osteoid osteomas has not been noted with these lesions.

Histology

Benign osteblastomas are considerably larger than the usual osteoid osteoma being more than 1 centimetre in diameter and often several times as large as this. Histologically however (Figs 140-142), they are made up of the same components as the osteoid osteoma, consisting of vascular spindle celled osteoblastic tissue with numerous trabeculae of osteoid tissue and calcified bone.

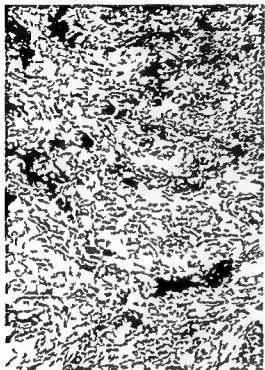


FIG 141 —Tissue from another case of benign osteoblastoma showing spindle celled osteoblastic tissue with areas of osteoid matrix and some abnormal calcified bone. Female aged 14 years with tumour of third lumbar vertebra. Golding and Sissons, Case 2, 1954 (x 63) (By courtesy of Editor *J. Bone Jt. Surg.*)



FIG 142 —Field from another case of benign osteoblastoma showing cellular osteoblastic tissue with prominent trabeculae of non lamellar bone. On superficial examination this type of tissue occasionally suggests a diagnosis of osteosarcoma (x 70)

Scattered osteoclast giant cells are present and may be conspicuous. In contrast to osteoid osteoma they do not appear to show a central region of calcified bone tissue such material being irregularly distributed throughout the lesion. The histological evidence indicates a relationship between benign osteoblastoma and osteoid osteoma although the differences in distribution of the two lesions in their size and in the reaction of the surrounding bone tissue appear to justify their recognition as distinct entities.

Although the presence of active osteoblastic bone formation may suggest osteosarcoma particularly in a small biopsy fragment real histological evidence of

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consequently find important application in connection with spinal tumours. The publications of Coley and his colleagues (Coley, Sharp and Ellis, 1931, Snyder and Coley 1945) established the value of this technique, and more recent accounts of its use have been published by Ottolenghi (1955) and by Schajowicz (1955).

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CHAPTER 11

RADIOLOGY

L. S. CARSTAIRS

GENERAL

THE radiographic image of a vertebral body is the sum of the shadows cast by the calcified trabeculae and the cortex. If the x rays used are of low penetrating power or the quantity is too small insufficient will pass through the vertebral body to produce blackening of the photographic emulsion and the shadow will appear to be homogeneously dense. With increasing penetration or quantity, or both some x rays will emerge from the opposite side of the body and will produce a pattern within its shadow corresponding to the main lines of blockage or absorption of the x ray beam. A 2 millimetre slice may contain several hundred trabeculae but if it is radiographed "end on" only one or two separate line shadows may be visible. This trabecular pattern cannot therefore represent all the bone elements that have produced it, but it does give an indication of their type and general arrangement. When there is a thin calcium containing sheet it will cast a dense linear shadow if a large enough part lies in the same plane as the x ray beam.

The vertebral body is sharply defined by its cortex. In lateral radiographs its shadow is of uniform density from front to back. This is unexpected because the body varies regularly in thickness the anterior surface being curved. It would be expected therefore, that the anterior portion would be less dense than the middle. The reason appears to lie in the cortex which forms a cylindrical shell. The x rays impinging at right angles to the shell in the middle of the body traverse only the thickness of the shell but in the curved anterior section they traverse the shell at increasingly oblique angles so that a corresponding increase in their absorption occurs. In dried preparations the fade off at the anterior margin can be seen but this is lost in vivo although it may be just visible in some patients in the cervical and thoracic spines where the physical conditions for contrast are better than in the lumbar spine. The part played by the cortex in producing the shadow of the vertebral body is greater than its thickness would suggest because of its compact structure.

Calcium content

An estimation of calcium content is necessary when considering generalized porosis or sclerosis. At the present time this direct estimation is subjective and is made by reference to known normals. The relationship of density to quality and quantity of x rays has already been mentioned. Other technical factors are involved including the size of the patient. Within limits it is possible to produce a dense shadow from a relatively porotic vertebra and a poor shadow from a dense vertebra. Obviously it would be desirable to have some objective method of comparison with the average. In the case of the hand this can be done by

GENERALIZED POROSIS

radiographing on one film the subject's hand and a control specimen, both being immersed in the same depth of water which removes the soft tissue shadows except fat (Jackson, 1951). More commonly a finger in a fluid filled plastic box is used the density of the box and its contained fluid corresponding to the soft tissues of a finger. Such methods or adaptations of them are feasible in the case of the spine but the practical difficulties are self evident. Apart therefore from changes in the shape of the vertebral bodies secondary to altered calcium content estimation of porosis or sclerosis is liable to considerable observational error.

GENERALIZED POROSIS

Introductory

The diagnosis of generalized vertebral porosis is based on two appearances (a) an estimate of calcium content, and (b) an alteration in the shape of the vertebral body. In porotic conditions the trabeculae become either fewer or less well calcified. The cortex appears to be less affected. In the lateral film of a normal lumbar vertebra the cortex on the anterior margin is usually not visible as a separate structure, the sum of the densities of the underlying trabeculae and the antero-lateral portions of the cortex being approximately equal to the density of the anterior cortex. In the antero-posterior films however, the cortex can often be clearly seen at the lateral margins. There is probably a physical explanation for this because on section of the vertebral body no alteration in the cortical thickness or the density of the trabeculae at the anterior and lateral portions is visible. In porotic conditions the sum of the densities of the trabeculae is reduced and the anterior cortex becomes visible as a thin pencilled line. It may be useful to compare the density of a lumbar body in a lateral film with the density of the soft tissues anterior to it. In severe porosis these two densities become almost equal.

Deformation

Reduction in calcium content results in structural weakness and this may be shown in two ways (a) fracture from minimal trauma and (b) a progressive collapse. The fracture is usually central producing a concavity in the upper or lower surface of the body. It may be limited to this or be associated with collapse of the peripheral cortex which usually occurs a short distance below the antero-superior angle, a small step being formed. Immediately after such a fracture the film will show some additional densities at the fracture site due to compaction of trabeculae and this may assist in differentiating the condition from collapse due to a destructive metastasis. Subsequently, the additional density may increase from callus formation but will disappear as remodelling occurs. The degree of internal damage may be greater than the radiograph indicates, and this can sometimes be supposed if there is a bulge in the anterior surface of the body. In such cases further collapse may occur before remodelling and consolidation. In those cases presenting as a slowly progressive collapse the underlying pathology may be either multiple minute trabecular fractures or a failure of the locally specific response to mechanical stress which in health, maintains the shape

of individual bones. The body becomes concave on its upper and lower surfaces (fish vertebra) There are two appearances which must be noted in relation to these changes. In the thoracic spine a lateral film may appear to show multiple concavities on the disc surfaces and this may be because the projection is not truly lateral. In some individuals the disc surfaces may, in health, be markedly concave. This concavity is usually greatest posterior to the mid point of the vertebral body, and alters to a convexity in the anterior third of the disc surface. It is very common in children in the lumbar region and pronounced examples are sometimes seen in adults. Such bodies are of course not exempt from porosis.

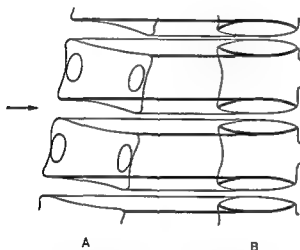


FIG 143 —(A) Vertebrae not correctly aligned with x ray beam. (B) Resulting image shows apparent biconcave disc surfaces.



FIG 144 —Markedly biconcave disc surfaces in young adult. Normal.

Disc changes

The deformation of the body causes the disc space to become convex on one or both surfaces. This implies that the intervertebral disc has undergone an increase in volume. In many cases of multiple vertebral body collapse this increase in total volume of the discs is quite indisputable. The primary lesion need not be a porosis nor is every porosis accompanied by disc swelling. In some cases it would seem that the disc change is a positive one and not merely secondary to the altered shape of the body. Several problems exist which require solution. Is the disc swelling due to a local change in vascular supply and diffusion rates or is it part of the general process that causes a particular type of porosis? Is it possible that in some patients swelling of the disc occurs before any evidence of body collapse is seen? One remarkable feature in some cases of porosis is that the lesion is reversible. This has been well illustrated by Wang and Robbins (1956) in a case of Cushing's syndrome. Severe porosis with collapse and disc swelling was present but three years after pituitary irradiation the vertebral bodies had almost regained their normal size and the disc space had returned to normal. Disc swelling can also occur in the diseased disc provided of course



FIG. 145—Disc swelling
 (a) Osteoporosis in an elderly woman with considerable swelling of disc substance which contains numerous calcium deposits (tracing) (b) Sclerotic deposits from a small bronchial neoplasm. Despite their increased calcium content the vertebral bodies are collapsing and there is considerable disc swelling (female)





FIG 146—Two chicken bones. Their density and trabecular pattern are comparable. The bone on the right has been kept at a high temperature and will crumble at the slightest touch.

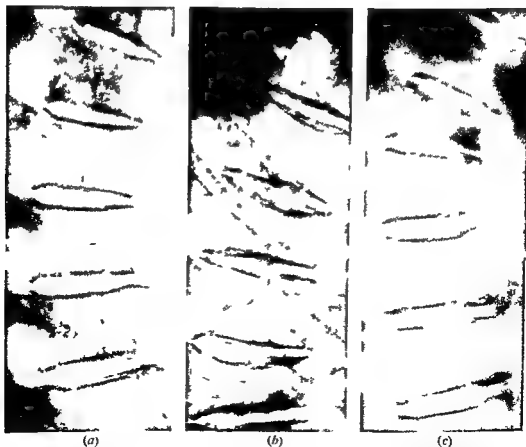


FIG 147—Porosis. (a) Carcinomatosis. Note vertical striation (Breast). (b) Osteomalacia steatorrhoea. Woman aged 38 years. (c) Carcinomatosis (breast). Uniform porosis. One collapsed body.



FIG 147 cont—(d) Reticulum-cell sarcoma, generalized (e) Lymphatic leukaemia
(f) Fragilitas ossium Woman aged 38 years

that anchorage by lateral osteophytic bridges is not present. In the case illustrated the discs are excessively large but there are also central areas of calcification which one would normally associate with long standing degenerative changes in the nucleus pulposus.

Density and structural strength

The calcium in the bony trabeculae is in several forms and the ratio of the forms may vary. X ray diffraction studies show that the calcium composition varies in different conditions (Cameron 1952). The radiologically demonstrable calcium content is by no means a certain indication of the structural strength. To take an extreme example Fig 146 shows radiographs of two chicken bones. They appear to be identical. On the right the femur has been heated and will crumble at the slightest touch. On the left the bone is fresh and untreated and is of normal strength. In marble bone disease the calcium content is high but the bones tend to be brittle. In rickets the calcium content is low but the bones are not fragile although they deform easily. In osteogenesis imperfecta the bones

may be of comparable density and cortical thickness to normal bone and yet undergo repeated fractures. With neoplastic invasion the bones may appear normal in structure, but the bony trabeculae have lost their normal cellular elements and the body may collapse at any time. In radiation necrosis unassociated with infection there is usually no alteration in the radiological appearances until the bone breaks.

It would seem that there are several factors (a) the composition of the calcium salts in the trabeculae, (b) the manner in which these calcium salts are laid down in the physical sense and (c) the nature and quantity of the cellular elements associated with the trabeculae.

This may seem to be far removed from the sphere of radio diagnosis but the radiologist is often puzzled by the discrepancy in behaviour of spines of comparable density in elderly patients.

Aetiology

So far the term *porosis* has been deliberately used in order to avoid confusion with the histological terms *osteoporosis* and *osteomalacia*. Both conditions produce the radiological picture of a reduction in density and in most cases it is not possible to differentiate between them in the vertebral column. These two pathological conditions are the centre of considerable interest at the present time, but in practice the first condition to be considered in a general spinal *porosis* is neoplastic infiltration—secondary carcinoma, myelomatosis and the leukaemias. Electroconvulsive therapy can give rise to multiple collapses particularly in the thoracic spine which mimic very closely the type of collapse seen in *porosis*. For example in one patient a chest radiograph taken after delivery following a normal pregnancy showed in the lateral film six partially collapsed upper thoracic vertebrae in which the upper surfaces of the vertebral bodies were concave. The possibility of a pregnancy osteoporosis was considered but the patient was subsequently found to have concealed the fact that she had had electroconvulsive therapy 5 years before.

Newbury and Eiter (1955) carried out a controlled observation in 106 patients receiving electroconvulsive therapy. The patients' spines were examined under standard radiological conditions before and after treatment. Twenty three vertebral body fractures were discovered. 13 of the patients had had curare and in these only 2 fractures were seen. None of the patients with fractures complained of pain.

Osteomalacia

The aetiological factors in *osteomalacia* have been dealt with elsewhere (Chapter 5). In some cases of long standing *osteomalacia* the spinal column presents a coarse vertical striation. This can occur in carcinomatosis and is also seen in simple or idiopathic *osteoporosis*. Radiological examination of the remainder of the skeleton in *osteomalacia* will often reveal pseudofractures. The onset of secondary hyperparathyroidism may be shown by cortical absorptions in the shafts of the phalanges of the fingers and toes and more rarely by lesions in the remainder of the skeleton. (It may be noted that Schinz and his colleagues

(1952) classified the skeletal changes in all osteomalacias with the exception of rickets due to avitaminosis D as secondary hyperparathyroidism)

In assessing progress in osteomalacia radiologically the two main features are (a) healing of pseudofractures, and (b) an increase in density of the vertebral body

Idiopathic osteoporosis

It has already been pointed out that in the spine differentiation from osteomalacia is often impossible. In osteoporosis the radiological findings are a decrease

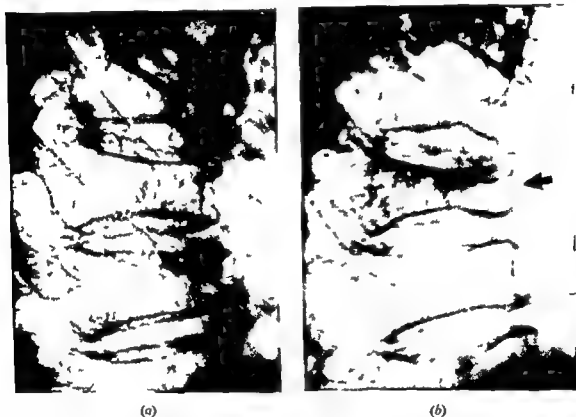


FIG 148 —Idiopathic osteoporosis in elderly woman (a) 22.9.56 Multiple collapsed thoracic vertebrae. Unchanged for 7 years (b) 31.1.57 Recent traumatic collapse shown by band of increased density

in density with structural weakness and consequent collapse, the healing of fractures occurs normally. The commonest single type of osteoporosis is that occurring in women past middle age. The term menopausal osteoporosis implies for the menopause a degree of aetiological importance that may not be justified (Donaldson and Nassim 1954). In general, the process is considered to be one of normal aging which in some patients for reasons unknown is very much more pronounced than in others. In children the possibility of a late form of osteogenesis imperfecta should be borne in mind while in adolescence apart from other causes of osteomalacia, the rare vitamin D resistant rickets of late onset must be considered. Prolonged immobilization of any part of the body

produces structural changes in bone. The spine is no exception and in children it may be a very marked feature of any prolonged recumbency (Halvorsen, 1954)

Cushing's syndrome and cortisone induced porosis

The severe porosis with vertebral collapse seen in Cushing's syndrome with the occasional reversibility of these changes has already been mentioned. Multiple rib fractures are common and callus formation is good even abundant. The degree of bone change is of course, related to the severity and duration of the disease. In cases investigated early, no rib fractures and no demonstrable porosis may be found. With the increasing therapeutic use of ACTH, cortisone and allied substances numerous examples of vertebral porosis have been recorded. Luder (1954) surveyed a series of such conditions in children occurring within 6 weeks to 1 year of the commencement of cortisone therapy (total dosage from 5.32 to 24.75 grammes of cortisone). The radiological picture resembled that of Cushing's syndrome. Adult asthmatics on baseline doses of cortisone and leading a fairly active life are particularly liable to isolated collapse of a vertebral body. In cortisone type fractures pain is generally not severe.

Other hormonal poroses

Excessive doses of thyroid hormone or even prolonged dosage with small quantities of thyroid can produce porosis. Severe porosis has been reported in testicular atrophy (Schinz and his colleagues, 1952) but considerable basic investigation of a large series of male castrates is required. Further differentiation is also needed in cases of hypopituitarism with hypogonadism and in cretinism. In these conditions it is difficult to say whether the deformation of the body is primarily a developmental fault or whether osteoporosis has contributed. In hyperparathyroidism the porotic changes are due to a combination of osteitis fibrosa and osteoporosis. The cause of pregnancy osteoporosis (segregated from deficiency osteomalacia) may be adrenocortical over activity during pregnancy (Nordin and Roper, 1955).

Congenital abnormalities

In some congenital abnormalities the spine undoubtedly appears to be porotic. This must be borne in mind in differential diagnosis particularly in children. In osteogenesis imperfecta there is often a spinal porosis and this may be severe with flattened bodies and biconvex discs. It may be difficult to differentiate some of these cases from osteomalacia at a single radiological examination. In general however the condition of the epiphyses and metaphyses will assist. Similar difficulties may be encountered in achondroplasia in children.

Neoplastic and haemopoietic

The general porosis of early but widespread metastases and the progressive porosis of leukaemia and the more chronic lipid reticulososes such as Gaucher's disease are well known. In carcinomatosis and myelomatosis the process is seldom completely uniform and areas of localized destruction can usually be found. In sickle cell anaemia in children generalized porosis with biconvexity of the discs has been reported but massive collapse is rare. In general the condition is subse-

LOCALIZED POROSIS

Capillary thromboses are a feature of the disease, and what was presumed to be infarction of two lumbar vertebrae was reported by Legint and Ball (1948). In Cooley's anaemia there is porosis of the spine in childhood followed by coarse reticulation in later life.

LOCALIZED POROSIS

In the spine localized porosis is a test for both radiographer and radiologist. Radiological examination is often shown to be misleading by the course of events and an appreciation of the limitations of radiography is desirable.

The formation of the image has already been discussed. The radiation required is of fairly high penetrating power and, apart from cortical densities, the difference in density of the body shadow and the surrounding 12 inches of soft tissues is not great. The structures within the vertebral body other than those containing calcium are of similar density to these surrounding tissues. When destruction of bony trabeculae occurs within the vertebra the space left is filled by soft tissue; the amount of calcium decreases but the amount of soft tissue increases.

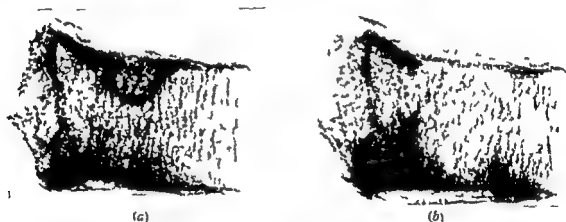


FIG. 149.—Radiographs (in water) of a vertebral body: (a) intact; (b) after a 1.5 centimetre hole has been made through the centre of the body. Apart from the removal of the Schmorl's node the hole is just visible.

Ardran (1951) showed that 3 millimetre and 14 millimetre holes drilled axially through a vertebral body are clearly visible if a contrasting substance such as air fills the spaces; but if the body is filled with water the 3 millimetre hole cannot be seen and the 14 millimetre hole is only just visible. Knutsson (1953) re-emphasized this and compared the efficiency of plain radiography and tomography. Chassin (1928) and Bohmig and Prevot (1931) had previously pointed out that large defects up to 15 millimetres in diameter were below the limits of visibility on plain radiography.

These experiments are easy and quick to perform and they provide a most salutary lesson in the limitations of radiography. In practice however it is well known that minute defects in the cortex, if seen tangentially, are clearly visible. Knutsson stressed this and amplified it showing that the defect in the cortex made by removing a layer 2 millimetres in thickness from the side of a vertebral body is clearly visible. These remarks refer to localized areas of total deficiency

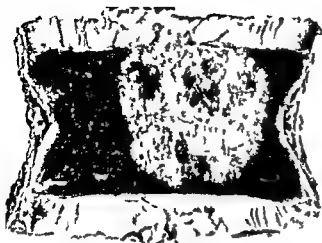


FIG 150 —The top photograph is of the cut surface of the vertebral body showing a large circular metastasis. Below is a radiograph of a 2 millimetre slice from this surface. The trabeculae are intact.



(a)



(b)

FIG 151 —(a) Collapsed body. No primary neoplasm detected clinically. Tomography shows areas of destruction and sclerosis. At necropsy there were wide spread metastases from a small bronchial neoplasm. (b) Woman aged 83 years. Acute tuberculous destruction. Miliary spread was shown at necropsy.

GENERALIZED SCLEROSIS

of trabeculae or cortex, but such lesions are not by any means, in most cases, encountered in clinical practice. Scattered small foci of destruction and diffuse intertrabecular infiltration without bone destruction often occur and usually escape detection. In the former there may be only the impression of porosis and in some cases a minimal loss of definition of the interior cortex, in the latter, no abnormality is visible (Fig 150). For a time the trabeculae maintain their strength but with the loss of normal cellular associations they become weak and the entire body may suddenly collapse. Alternatively, the abnormal cellular infiltration produces a diffuse thinning of the trabeculae with or without small areas of destruction and the picture is that of porosis undergoing slow collapse or subject to sudden collapse after minimal trauma. This is one of the commoner findings in myelomatosis. In the infective conditions, hyperaemic decalcification, in addition to trabecular destruction and fracture takes place. A localized hyperaemic decalcification also occurs in some cases of trauma but this is probably secondary to multiple fine trabecular fractures.

GENERALIZED SCLEROSIS

A true uniform sclerosis of the entire vertebral column is a rarity, in all conditions there is usually some patchiness. The most uniform densities are produced by (a) long standing widespread sclerotic deposits particularly from the prostate (b) chronic fluorosis which is usually associated with marked osteophytosis and ligamentous ossification but cases of fluorosis have been reported in which these associated changes are either absent or minimal (Linsman and McMurray, 1943. Largent Bouard and Heyroth 1951) and (c) marble bone disease in which the sclerotic bone is mainly epiphyseal producing dense bands at the upper and lower margins of the bodies and occasionally central densities corresponding to the primary ossific centres. In these conditions the abnormality is obvious. Marginal increases in density are more difficult to assess. In some young adults the density of the skeleton, and particularly the vertebral column, is slightly more than one would expect but this appears to be of no significance and is only an expression of the individual variations in size and number of the bony trabeculae in health. In 16 cases of idiopathic hypercalcaemia of infants reported by Creery and Neill (1954) the vertebral column was normal and there was no nephrocalcinosis. Thirteen however showed dense bands in the metacarpals radius ulna, tibia or femur, reminiscent of the dense bands of lead poisoning. The prognosis was good in all cases and in 4 followed to recovery the dense bands disappeared. In a case reported by Lowe and his colleagues (1954) there was a uniform density throughout the skeleton including the vertebral column with very dense bands at the metaphyses.

Cases of osteomalacia with secondary hyperparathyroidism from chronic renal failure with glomerular damage and urea retention have occasionally presented an unusual density of the bones (Crawford and his colleagues 1956). Radiologically there are dense bands in the upper and lower portions of the vertebral body similar in type but not in density to those of marble bone disease. The density is an unexpected finding of unknown cause. During experiments concerned with the effect of parathormone injections on the growing skeleton (as part of a study related to the clinical use of parathormone in lead poisoning) Bauer Aub and Albright (1929) found that in young growing rats on a high calcium diet 885

units of parathormone given over 110 days produced bones that were shorter but radiographically denser than normal. This ran counter to their results with other animals and they were not able to offer an explanation. Selye (1932) carried out a series of experiments with albino rats using varying dosages of parathormone. He concluded (a) that massive doses produced osteitis fibrosa followed by the supervention of osteoblastic activity and suggested that Bauer's results could be explained by this. (b) that very small doses directly stimulated new bone formation without a preliminary stage of osteitis fibrosa. (c) that a single dose of 10 units of parathormone could stimulate new bone formation, this conclusion being reached by killing rats at intervals during 48 hours. Selye regarded the changes produced in the rats as comparable with those of marble bone disease.

In all such cases of dense bands in the metaphyses it is essential to exclude the possibility of intoxication by a heavy metal, particularly lead in young children.

In cases of generalized sclerosis produced by long standing prostatic deposits the pelvis and upper femora will usually reveal areas where the sclerosis is patchy. Generalized sclerosis in cases of breast carcinoma are most common after hormonal therapy or pituitary ablation and the history is all important. Although uniform density without expansion can be present in Paget's disease, it is uncommon and in general the enlargement of the vertebral bodies, the coarsening of the trabecular pattern, particularly in the peripheral portion of the body, and the tendency to flattening will be obvious. Generalized patchy sclerosis occurs in sickle cell anaemia and Cooley's anaemia, acquired haemolytic anaemia and the rare myelosclerotic anaemia.

LOCALIZED SCLEROSIS

The cause of the sclerosis sometimes occurring in bone subject to abnormal mechanical stress is not understood. Typical examples are often seen in the spine commonly in the antero-inferior or antero-superior segments of vertebral bodies adjacent to disc degeneration and also in ankylosing spondylitis. In this condition the general rigidity of the spine is sometimes partly compensated for by localized hypermobility at one or more disc spaces. When this occurs localized sclerosis of the type mentioned above may occur and is in strong contrast with the generalized porosis which is a feature of the disease. Isolated sharply defined sclerotic lesions reaching to the cortex are sometimes seen and remain unaltered over several years; they are given the name 'compact bone island', 'enostosis' or 'calcified medullary defect'. Martincic (1952) reported a case of osteopoikilie in which the thoracic and lumbar sections of the spine were affected. In the neuro-pathic spine some of the vertebral bodies, particularly in the lumbar region, appear to be dense with an ill-defined cortex and have a peculiar 'toppling' appearance associated with multiple disc degenerations. There may also be areas of porosis and apparent fragmentation of the disc surfaces with lateral collapse.

The solitary sclerosed vertebra usually referred to as an 'ivory vertebra' is a special problem. It may be due to lymphadenoma, Paget's disease or sclerotic metastasis; occasionally examples are encountered in which no cause is found and some vascular disturbance is suggested. On the analogy of avascular necrosis at other sites in the skeleton one would expect in such a case some collapse of

ANKYLOSIS

the body to be present caused by structural weakness or that the condition would follow the pattern of delayed collapse after injury (Kuemmell's disease). In this condition there is usually some rarefaction of the body before collapse but sclerosis can occur and the association of bone injury with infective lesions should be borne in mind as in a case reported by Schinz and his colleagues (1952) in which a purulent pyelomyelitis was present at the level of the collapsed and sclerosed body. Meltzer (1947) reported a case in which a lipoma in the spinal cord was associated with sclerosis of the posterior part of the bodies of the sixth and seventh cervical vertebrae the laminae and the spinous processes, at operation and on biopsy there was no evidence of bone infection or metastases. Pygott and Scott (1954) reported a case of leontiasis ossea of the Virchow type in which in addition to the changes in the skull the facial bones the ribs and one tibia there was a dense sclerosis of four thoracic vertebrae.

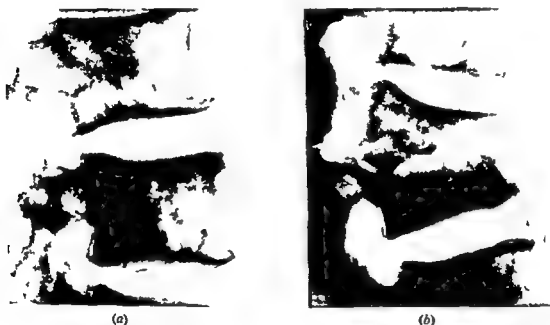


FIG 152—Lymphadenoma. (a) Sclerosis in body. Male patient presented with back pain and solitary round shadow in right lung. (b) Collapse with disc destruction. Rare. Post mortem specimen.

ANKYLOSIS

Congenital fusion of the vertebrae is reasonably easy to recognize because the trabecular pattern is normal and there is usually no loss of substance. In juvenile rheumatoid arthritis with spinal involvement fusion of the posterior joints without ossification of intervertebral ligaments may be mistaken for a congenital lesion (Barkin Stillman and Potter 1955). Bony ankylosis is commonly infective but in the assessment of long standing fusions the possibility of a post traumatic synostosis without infection should be remembered. Secondary infection however does occur in traumatic haematomas and unless the clinical history is available differential diagnosis is difficult. Ankylosis has also been reported in a rapidly advancing case of Paget's disease of the cervical spine treated by high calcium diet (Stein Stein and Beller 1955). In elderly patients who are not

suffering from ankylosing spondylitis, fusion of anterior osteophytes is common and marked ligamentous ossification with little or no protruding osteophytosis also occurs (Smith Pugh and Polley, 1955)

The nature of ankylosing spondylitis is unknown. Romanus (1953) after a most careful and searching investigation, concluded that an infective element was important in most cases. This hypothesis has not gained general acceptance in Great Britain. Clinically and radiologically the disease can be differentiated from rheumatoid arthritis. Gibson (1957) concluded from hereditary and genetic factors, serology and clinical pathology that although the two diseases are of the same family they are no more related than acute rheumatism and polyarteritis nodosa or rheumatoid arthritis and scleroderma. The clinical course and physical signs are of paramount importance in early diagnosis. Radiologically, the classical appearances of the active disease in the sacro iliac joints in adults is unmistakable but difficulty may be encountered during adolescence at the times of appearance of the plate epiphyses and, at an earlier age in differentiating from tuberculous lesions. There are, however, lesions in the sacro iliac joints unassociated with pathognomonic vertebral changes, consisting of sclerosis and erosion of the iliac surfaces with minimal changes in the sacral surfaces in which little alteration occurs over several years and clinically the main symptoms are low back pain rather than decreased spinal mobility. These may be cases of aborted ankylosing spondylitis or a low grade non specific arthritis of unknown aetiology. Since radiotherapy is an established form of treatment for ankylosing spondylitis it is important that such cases should not be misdiagnosed and re assessment after a period of observation may be desirable.

PRESSURE EROSIONS

Erosion of the anterior and lateral surfaces of the bodies may occur in aneurysm glandular neoplasm abscess and benign neoplasm. Widening of the interpedicular space with spinal canal tumours is well recognized. Difficulty may be encountered in the thoracic region in children where some variation in the radiological appearances is common. A case of localized widening and apparent thinning of

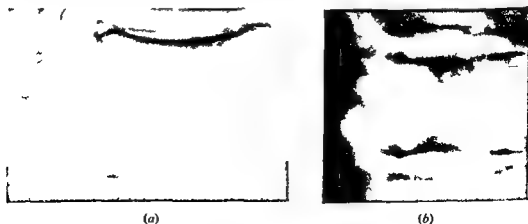


FIG 153—Concave posterior surface. (a) Neurofibromatosis of cauda equina. (b) Metastasis. New growth in breast.

the pedicles of T 12 was reported by Jefferson (1955). Operation revealed a normal canal and contents. The lesion was a congenital abnormality. Widening of the exit foramen with pedicle erosion by neurofibromas is also well recognized, but this diagnosis must be made with caution in the thoracic region because the plain radiographic appearances can be exactly mimicked by thoracic meningocele. The soft tissue paravertebral mass may be found to change its shape with alterations in posture. Myelography is conclusive and should not be omitted. Raison (1956) reviewed 26 previously reported cases and presented two additional examples. A further point of confusion arises from the very frequent association of von Recklinghausen's disease with a thoracic meningocele (17 out of 28 cases). In the lumbosacral area extradural cysts may produce erosions of the pedicles and body and neural arches identical with those seen in neurofibromatosis. The subject has been reviewed by Schurr (1955). All degrees of anterior sacral meningocele are encountered from a shallow erosion on the anterior surface of the sacral canal to a large defect with a soft tissue mass lying between the sacrum and the rectum.

THE INTERVERTEBRAL DISC

The healthy disc cannot be seen on plain radiography because its density is like that of the surrounding tissues. The size in relation to that of other discs can be assessed and films taken in lateral flexion, extension and forward flexion can give some idea whether or not the disc responds normally to spinal movement. Radiologists have always been interested in the physical properties of the nucleus pulposus and the annulus and many divergent views have been expressed ranging from the concept of tonus in the annulus (Roberts 1944) to the idea of turgor in the nucleus pulposus (Schmorl 1931). It seems certain that the healthy disc maintains its elasticity because of the physical properties of collagen and the nature of gel structure. Exposure of an intervertebral disc to the air very rapidly produces drying with a remarkable change in size and consistency illustrating how dependent the disc is on transudation of fluid from its environment. Naylor, Happey and Macrae (1954) have investigated by x-ray crystallography, the composition of the annulus and nucleus in material taken from subjects ranging from 24 to 78 years under standard conditions of drying and by frozen section. They concluded that after the third decade (a) alteration in the orientation and decreased mobility of the collagen fibrils occur in the annulus and (b) that increased orientation and ultimate crystallization causing a loss of gel structure occurs in the nucleus. Both conditions would result in a loss of elasticity. Post-traumatic and degenerative changes in the disc have been fully described and illustrated by Harris and McNab (1954). Radiographically, however, only two conditions are demonstrable: these are vacuum phenomena and disc calcification.

Vacuum phenomena

All body fluids contain gases in solution and when the physical conditions allow these gases can be pulled out of solution by decreasing the pressure to which the fluid is normally subject.

The phenomenon is very common and several hundred examples are readily collected in routine hospital practice. Two types are seen (a) linear streaks of gas in the disc space extending through the whole or part of its width and

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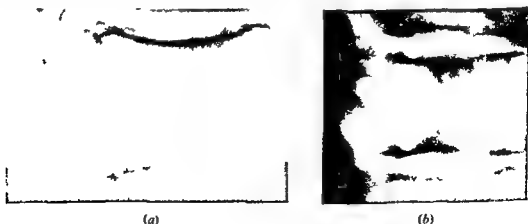


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Vacuum phenomena

All body fluids contain gases in solution and when the physical conditions allow these gases can be pulled out of solution by decreasing the pressure to which the fluid is normally subject.

The phenomenon is very common and several hundred examples are readily collected in routine hospital practice. Two types are seen: (a) linear streaks of gas in the disc space extending through the whole or part of its width and

(b) minute spherical collections situated at the anterior or antero lateral margins of the body in the disc space. The linear streaks (nucleus pulposus type) are generally not affected by alteration in posture and the appearance may remain unchanged for many years. It is suggested that this type is produced by cracking and then desiccation of the central disc substance. The vertebral bodies approximate but complete apposition is prevented by the residual parts of the annulus. The desiccating process continues and for some reason unknown the space is filled by gas and not by fluid. The small marginal spherical shadows (annulus type) do appear to be affected by changes in posture and by traction thus behaving like the gas shadows in joints. It is supposed that they lie in small cracks in the annulus and that these cracks are degenerative and precede the formation of marginal osteophytes. The writer is not aware of any record of necropsy findings in a case in which a vacuum phenomenon had been described during life.

The importance of the condition lies in (a) positive proof of a disc abnormality, and (b) exclusion of tuberculous infection if the disc space is narrowed.

Disc calcification

The deposition of calcium in degenerating tissue is by itself asymptomatic apart from loss of flexibility. The physiochemical basis is poorly understood. Calcification also appears to occur rapidly as in tendinous calcification in the region of the joints notably the shoulder, elbow and hip. The deposit may be in a state of increased tension and may cause pain either by distension or by perforation of the surrounding tissue and spread into the associated bursae or tissue planes. These two types of calcification also occur in the intervertebral disc. The first the degenerative type is very common while the second which one might call the active type is rare. Sandstrom (1951) preferred to classify them as permanent and transitory.

The degenerative type follows the distribution of the vacuum phenomenon that is in the annulus anteriorly and antero laterally and in the nucleus pulposus. It is very common and is seen at all levels from the cervical to the sacrococcygeal articulation. It is probably most common in the mid and lower thoracic and upper lumbar regions. The shape of the calcium deposition also parallels that of the vacuum phenomenon. In some cases which appear to be locally asymptomatic the calcium takes the form of one or two spherical deposits in the nuclear region. Occasionally it forms a sharply defined oval opacity within the disc space. Multiple disc calcifications are common in patients in whom spinal mobility has been completely lost because of ankylosing spondylitis or advanced spondylosis with anterior vertebral fusion.

Examples of complete posterior extrusion of calcified disc substances have been reported and a well documented and illustrated example of this in the thoracic spine has been published by Williams (1954a). Logue (1952) reported that of 11 thoracic disc protrusions proved at operation there was disc calcification at the same level in 5.

The active or transitory type of disc calcification is of considerable clinical interest. Since a report of one case by Baron (1924) further cases have been reported sporadically and there are now over 20 cases in the literature. Most occurred in boys but two reports (Sandstrom 1951) refer to adults past middle age. In all cases there was localized pain and tenderness with limitation of move-

ment. Associated malaise, fever and raised sedimentation rate have been observed. The calcification disappears completely or partially after a few months to several years.

Percher and Storrs (1956) reported two cases. In one, in a boy aged 4 years, there was severe pain of slow onset in the neck with anorexia. All investigations



(a)



(b)

FIG. 154—Disc calcification in woman. Known carcinoma in cervix. Pain in back. (a) 4.6.56. Diffuse disc calcification. (b) 22.11.56. Most of the calcium has been absorbed leaving a small central calcification. (c) 11.4.57. Collapsed vertebral body at same level due to metastasis. Generalized deposits.



(c)

were negative apart from a C3/4 disc calcification which disappeared in 4 months. Walker (1954) reported a case in a girl aged 10 years in whom massive spherical calcifications were present at T11/12/L1/2. The disc spaces were widened posteriorly and there was a pronounced concavity of the vertebral bodies surrounding the spherical calcifications. Lateral escape of the calcified material occurred at one level; at another the calcification had absorbed completely in 4 months.

Most had gone after 3 years and 10 years later all had disappeared, leaving some residual narrowing of the disc spaces. This case illustrates the true increase in volume which some calcifications appear to induce. In the adult with back pain and in whom a spherical or ovoid calcification is demonstrated the problem arises whether or not this calcification is clinically significant. Radiologically no assistance can be given but by analogy with the peri articular tendinous lesions already mentioned change in these calcifications would suggest that they were significant. A further problem that arises is whether or not swelling of an isolated disc can occur (without calcification) and whether this might not be clinically significant. That an increase in the volume of disc spaces can be produced by



FIG 155 —Alkaptonuria. Showing disc calcification, disc obliteration with fusion of bodies and disc degeneration with vacuum phenomena.

traction has been shown by Christie (1955). Five minutes after spinal traction with a force of 75 pounds in 2 volunteers there was an increase in depth of the L4/5 and L5/S1 disc spaces by 10-15 per cent.

Osteochondritis

There are difficulties in recognizing the early changes of this condition in the spine (Williams 1954b). The mode of development of the vertebral body and in particular the part played by the epiphyses in vertical growth does not appear to be the subject of full agreement. The detailed work of Lacapre, Drieux and Kriegel (1952) is excellently illustrated and should be consulted. Ferguson (1956)

recently suggested that the wedging which is usually attributed to damage and faulty growth of the epiphyses is in some cases due to collapse of the vertebral body around the anterior vascular groove. Severe osteochondritis with the development of a thoracic kyphosis is a crippling disease in middle age because of the secondary changes in the thoracic cage and lungs. It would seem that further research into its aetiology and pathology is still required.

METASTASES AND VASCULAR SUPPLY

It is uncommon for the spine to be directly involved by neoplastic spread but when it occurs it is probably most frequent in the upper thoracic region from peripheral bronchial carcinomas in the apex of the upper lobes. Metastases are blood borne and opinion differs about how they reach the vertebral bodies. The generally accepted view is that they pass from the tumour site as emboli into the venous system and then pass through the heart and lungs reaching the vertebral bodies as arterial emboli. In the case of the prostate there is some acceptance of the suggestion made by Batson (1940) that the metastases spread from the pelvic veins into the vertebral venous plexus. If the arterial embolus theory were usually correct most metastatic bone lesions would be expected to be accompanied by pulmonary metastases. It has been shown however that glass spheres varying in diameter from 10 to 500 microns injected into the pulmonary arterial system can be collected from the pulmonary venous system and that there are direct communications between these two systems (Tobin and Zariquey 1953). The greater part of the pulmonary circulation however does pass through the pulmonary capillaries so that this diversion of emboli from the pulmonary capillaries should occur only in a small proportion of cases. It may be that pulmonary tissues are in some instances a less favourable site than bone tissues for the development of metastases. Batson (1940) showed that injection of opaque medium into the pelvic venous system can fill the whole of the vertebral venous plexus. Franks (1953) confirmed this work in 15 subjects and was also able to demonstrate in one case tumour cells and barium in a prostatic vein and barium filled venous spaces in vertebrae affected by deposits in another. Batson also demonstrated that injection of opaque material into veins at the areola of the breast can extend into the cervical venous plexus, the superior longitudinal sinus and even the coracoid process and humeral heads on opposite sides of the body. All these experiments were performed in the cadaver. Lindbom (1955) confirmed these findings in so far as the pelvic veins are concerned during life. Seventy five female patients ranging in age from 20 to 75 years and all suffering from carcinoma of the uterus were subjected to venography by catheterization of both femoral veins to the level of the junction of the internal and external iliac veins. The inferior vena cava was obstructed by compressing the abdomen with a football bladder and 30 millilitres of contrast medium were injected into each catheter simultaneously. The opaque medium passed into the pre sacral plexus and thence into the internal and external vertebral plexuses. The internal plexus lies in the spinal canal, the external lies outside the canal between the spinous processes and paravertebrally. The opaque medium filled the vertebral venous plexus and contrast was sufficiently good in these cases to note the compression of posteriorly protruded discs on the internal plexus lying against the posterior longitudinal ligament.

Harris and Jones (1956) investigated the arterial supply to the cervical vertebral bodies and showed that the vertebral artery gives off a spinal branch in the intervertebral foramen which then divides into three further branches (1) to supply the cord, (2) to supply the laminae ligamentous flava and so on and (3) to an arterial plexus situated on the dorsal surface of the vertebral body. From this plexus arise (a) a single wide artery which enters the body dorsally and proceeds to its centre where it divides into a number of small branches which radiate out from this central point and (b) circumflex arteries which send small

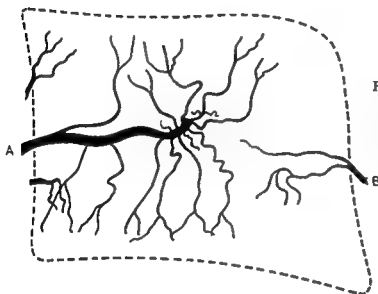


FIG 156—(A) Large artery arising from dorsal plexus (B) Small artery from circumflex plexus (By courtesy of *Journal of Bone and Joint Surgery*)

branches into the cortex around the circumference of the vertebral body. This type of distribution would suggest that large arterial emboli either neoplastic or infective would be held up in the middle of the body while small arterial emboli composed of bacteria would tend to reach the periphery of the body and its disc or lateral surfaces. The assumption made by Batson is that during increases in intra abdominal pressure the normal venous flow is reversed so that the portal and caval venous systems are by passed. In cases of cirrhosis Steiner, Sherlock and Turner (1957) showed on splenography that the vertebral veins participate in the alternative venous return system. While by passing of the portal and caval systems has been shown to occur it would still be necessary to suppose that a reversal of flow occurs in life within the vertebral body and it is evident that further work has still to be done to prove or disprove this point.

An attempt to utilize contrast injections for diagnostic purposes has been made by Lessmann and his colleagues (1955). Erhardt and Kniep (1943) suggested the infusion of opaque substances into the bone marrow as a diagnostic measure and this has been used for the demonstration of the venous system of the leg and pelvis. Lessmann injected the opaque medium into the spinous processes with three objectives (1) to investigate the patency of the azygos and hemiazygos

veins in diseases of the chest and mediastinum (2) to study the vertebral veins in normal and pathological circumstances and (3) to determine the feasibility of detecting isolated neoplastic bone involvement earlier than by conventional methods. (It may be noted in this respect that biopsy of a spinous process has been used in the assessment of the clinical condition of late carcinoma of the breast.) Eighty injections were made in 45 patients. In 10 cases of late bronchial neoplasm 5 veins were found not to fill and subsequently did fill after radiotherapy. In one case of generalized osteosclerotic metastases the internal venous plexus was normal. In one case there was a block of the internal venous plexus no abnormality was seen on plain radiography but on subsequent bone biopsy neoplastic invasion was found. Tori (1954) has also published excellent demonstrations of the internal plexus by the same method. The types of metastatic invasion and destruction have already been mentioned.

In the case of sclerotic metastases it is important to remember that as much neoplastic infiltration may be present in the intertrabecular spaces of radiologically normal bone as in the areas of sclerotic deposits.

RADIOLOGY AND LOW BACK PAIN WITHOUT SCIATIC RADIATION

Many suggestions have been made regarding the aetiology of the common forms of low back pain and a discussion would involve almost all the known pain producing conditions some of which are as yet hypothetical. Marginal osteophytosis is very frequent. Cartilage loss, sclerosis and osteophytosis occur in the posterior joints. Harris and McNab (1954) illustrated some of the other changes that occur and which might be aetiologicaly significant, for example alterations in the shape of the posterior facets caused by subluxation from disc degeneration, calcification in the capsule of the posterior joints, loose bodies in these joints, osteochondral fractures of the posterior joint surfaces, dense adhesions and the disorders of movement associated with disc degeneration. Some of these conditions may be shown by radiography but whether they are productive of symptoms in a particular patient must be a matter of clinical decision. For example Bistrom (1954) selected 151 subjects on the basis of (a) no history of back pain and (b) all engaged regularly in heavy manual labour or strenuous physical activity. Of these 8 had spondylolisthesis (including one complete forward dislocation), 5 had pseudospondylolisthesis, 30 had transitional lumbar vertebrae (of whom 10 had accessory joints which were associated with scoliosis in 5 cases). Roche and Rowe (1951) examined 4200 skeletons and found that 178 (4.2 per cent) had separate neural arches in the lumbar region (highest white males 6.4 per cent, lowest Negro females 1.1 per cent). In the case of disc degenerations Hirsch (1955) concluded that backache is not the result of the degeneration *per se* but is due to other factors including fracture of the vertebral plate and growth of granulation tissue into the disc after annular rupture. However by analogy with degenerative arthritis elsewhere the symptoms probably arise from changes in the surrounding soft tissues rather than in the components of a disordered joint. Fortunately in most patients the problem is not to make a completely accurate diagnosis but to provide a safe even if empiric method of treatment and if for this alone radiography is necessary to exclude infective and neoplastic processes before manipulation or traction.

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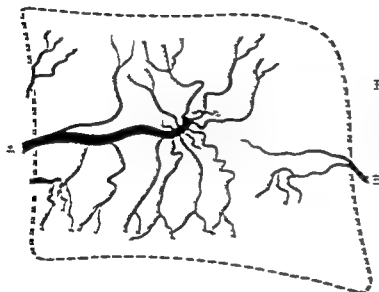


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An attempt to utilize contrast media for diagnostic purposes has been made by Lessmann and his colleagues (1945). Erhardt and Kneip (1943) suggested the infusion of opaque substances into the bone marrow as a diagnostic measure, and this has been used for the demonstration of the venous system of the leg and pelvis. Lessmann injected the opaque medium into the synovial processes with three objectives: (1) to investigate the patency of the arthro and hemiarthro

veins in diseases of the chest and mediastinum (2) to study the vertebral veins in normal and pathological circumstances and (3) to determine the feasibility of detecting isolated neoplastic bone involvement earlier than by conventional methods. (It may be noted in this respect that biopsy of a spinous process has been used in the assessment of the clinical condition of late carcinoma of the breast.) Eighty injections were made in 45 patients. In 10 cases of late bronchial neoplasm 5 veins were found not to fill and subsequently did fill after radiotherapy. In one case of generalized osteosclerotic metastases the internal venous plexus was normal. In one case there was a block of the internal venous plexus no abnormality was seen on plain radiography but on subsequent bone biopsy neoplastic invasion was found. Tori (1954) has also published excellent demonstrations of the internal plexus by the same method. The types of metastatic invasion and destruction have already been mentioned.

In the case of sclerotic metastases it is important to remember that as much neoplastic infiltration may be present in the intertrabecular spaces of radiologically normal bone as in the areas of sclerotic deposits.

RADIOLOGY AND LOW BACK PAIN WITHOUT SCIATIC RADIATION

Many suggestions have been made regarding the aetiology of the common forms of low back pain and a discussion would involve almost all the known pain producing conditions some of which are as yet hypothetical. Marginal osteophytosis is very frequent. Cartilage loss, sclerosis and osteophytosis occur in the posterior joints. Harris and McNab (1954) illustrated some of the other changes that occur and which might be aetiological significant for example alterations in the shape of the posterior facets caused by subluxation from disc degeneration, calcification in the capsule of the posterior joints, loose bodies in these joints, osteochondral fractures of the posterior joint surfaces, dense adhesions and the disorders of movement associated with disc degeneration. Some of these conditions may be shown by radiography but whether they are productive of symptoms in a particular patient must be a matter of clinical decision. For example, Bistrom (1954) selected 151 subjects on the basis of (a) no history of back pain and (b) all engaged regularly in heavy manual labour or strenuous physical activity. Of these 11 had spondylolisthesis (including one complete forward dislocation), 5 had pseudospondylolisthesis, 30 had transitional lumbar vertebrae (of whom 10 had accessory joints which were associated with scoliosis in 5 cases). Roche and Rowe (1951) examined 4 200 skeletons and found that 178 (4.2 per cent) had separate neural arches in the lumbar region (highest white males 6.4 per cent, lowest Negro females 1.1 per cent). In the case of disc degenerations Hirsch (1955) concluded that backache is not the result of the degeneration *per se* but is due to other factors including fracture of the vertebral plate and growth of granulation tissue into the disc after annular rupture. However, by analogy with degenerative arthritis elsewhere the symptoms probably arise from changes in the surrounding soft tissues rather than in the components of a disordered joint. Fortunately in most patients the problem is not to make a completely accurate diagnosis but to provide a safe even if empiric method of treatment and if for this alone radiography is necessary to exclude infective and neoplastic processes before manipulation or traction.

RADIOLOGY AND LOW BACK PAIN WITH SCIATIC RADIATION

Pain in the distribution of the lumbar and sacral nerves usually indicates root irritation but it would be well to remember conditions which can simulate this.

McNeur (1953) reported 6 patients treated for presumed disc lesions whose hips (5 cases) and the humerus (1 case) had not been examined radiologically and were the seat of bone tumours.

Bonney (1956) reported 9 patients with gluteal pain in whom a diagnosis had been made of disc protrusions or arthritic hips: they were shown by aortography to have either aortic or iliac occlusion. He commented that the pain occurred with exercise and was constant in character.

It has often been suggested that pain could be referred from a degenerated disc to the soft tissue structures of the lower back but Steindler (1954) concluded that this was improbable. He had, however, noted a few cases in which Novocain injection into a tender spot in the lower back temporarily abolished the local pain and its associated sciatic radiation suggesting that the latter was in these cases, a reflex phenomenon. The radiologist cannot on plain radiography, reach any conclusion on whether a protruded disc is present or not unless a protruded disc is calcified. He can state however that when a disc space is unequivocally smaller than normal there is some abnormality of the disc, but even here some clinicians would hold that selective muscular contraction or spasm can produce disc narrowing or deformation. This may be so in the cervical spine but it is unlikely in the lumbar spine. Schalimtzek (1954) concluded after studying 5 anaesthetized and curarized subjects that abnormal disc size associated with abnormal movement was not muscular in origin. The essential evidence of involvement of a particular nerve root is the presence of abnormal neurological signs. Physical examination supplemented by plain radiography is in many hands sufficient for full diagnosis and provides adequate information for laminectomy. There is no doubt, however, that additional radiological investigation is of value in excluding other conditions and in localizing the offending disc. These supplementary methods are (a) motion studies; (b) discography, and (c) myelography.

Motion studies

Motion studies of the lumbar spine have long been suggested as a supplement to plain radiography. Films are taken in lateral flexion, extension and forward flexion and abnormal restriction and abnormal mobility may be discovered. Schalimtzek (1954) has reviewed this subject and has included the results of his own investigations and on the basis of operation findings compared its efficiency favourably with myelography. Basically this method does not diagnose disc protrusion but only disc abnormality. The decision whether or not a disc abnormality is associated with a protrusion is again made on clinical grounds.

Discography

A large number of reports of annular rupture, disc protrusion and disc degeneration following accidental puncture and experimental puncture in animals is available and the introduction by Lindblom (1948, 1950, 1951) of the technique of discography was something of a surprise. It is asserted that the use of a very fine bore needle obviates the risk of annular damage. The method necessitates

the introduction of a diffusible water soluble opaque medium into the centre of the disc. The results have been of the greatest value in demonstrating normal and abnormal disc structure. On occasion they may be the only method of demonstrating lateral protrusion producing foraminal encroachment.

It is necessary to know at approximately which level a protruded disc is clinically suspected. This disc and the one above and below are then injected under radiographic or fluoroscopic control and films are taken. The findings have been fully described by Ehrlicher (1952) as follows. In young healthy subjects the opaque medium spreads to outline a globular nucleus. A lobulated nucleus may be found in adults and with large lobulations protrusion is unlikely. A simple branched nucleus with a central shadow predisposes to herniation. Multiple branching with a small central shadow indicates likely prolapse. A spread nucleus with no central core and many branches running in all directions indicates a fully degenerated disc which is unlikely to herniate. Ehrlicher used a lateral approach by passing the theca.

Friedman and Goldner (1955) reported the diagnostic results of 427 disc injections in 150 patients in 50 of whom myelography was also performed. They found that discography demonstrated 22 diseased discs where myelography had disclosed none. There was a high correlation between discography and operative findings. Wolkin Sachs and Hoke (1955) came to the same conclusion in a small series but reported only partly accurate findings in 7 and a false positive in 1 out of 27 cases. The complex physicochemical structure of the normal nucleus pulposus might be expected to react unfavourably to the introduction of a solution containing a compound of high molecular weight.

DeSeze and Leverneux (1952) reviewed 59 patients one year after discography. Of these 28 had had symptoms directly attributable to the procedure. Ten had had severe lumbar pain for a few days to one month. One had had severe abdominal pain associated with spread of the contrast medium under the anterior ligament. At operation a cavity containing necrotic tissue was found in the disc region in 1 patient. Twelve patients developed violent lumbar pain about 6 days afterwards with thoraco lumbar rigidity and in some there occurred changes in the vertebral disc surfaces simulating tuberculous infection. Three patients developed meningism they had small posterior spindles allowing the contrast to escape. Two patients had venous thromboses one with pulmonary infarction. The authors concluded that these conditions were not due to either infection or sensitivity but simply to the injection of a contrast medium into the disc and that no single make of contrast medium was at fault. Ten patients had residual backache and 13 developed disc degeneration where none was previously seen. Finally where there is any clinical doubt of a neoplastic lesion in the spinal canal myelography will be required and any suggestion of an infective process is a direct contraindication to discography.

Myelography

Three methods are available gas water soluble medium and oily contrast medium.

Gas myelography is not in common use in Great Britain although there is much to be said in its favour particularly for the younger patient. The technique is described by Lindgren (1952).

The use of water soluble contrast medium is attractive because by this method the components of the cauda equina can be visualized throughout their length. The method requires spinal anaesthesia. Not all types of contrast media can be used and fatalities have been reported from the accidental use of the wrong material. The complication rate is high and the opaque medium must be confined to the lumbar region. For these reasons it has not gained wide acceptance. The oily medium Myodil (Pantopaque) is virtually non toxic it is gradually, if slowly absorbed and although for medico legal reasons it should ideally be withdrawn in many centres this is not done. Indeed unless the needle is left *in situ* during the examination the additional puncture may itself produce complications as in the case reported by Keats (1956) in which the medium was seen to disappear rapidly and form emboli in the lungs violent and uncontrollable coughing occurred but there were no severe symptoms and recovery was complete. To leave the opaque medium *in situ* has the advantage of allowing re examination at intervals and assessment of the results of operation. The method has limitations (Reid and Tutton 1951, McCarthy and Lane 1955) both in the diagnosis of disc protrusion and of other space occupying lesions in the spinal canal.

It is not possible to enter into a description of myelographic technique or interpretation but some selected observations may be of interest. One difficulty lies in the interpretation of uniform slight posterior bulging of the disc as shown in the lateral films. Generally this may be considered to be physiological particularly with the patient standing and in the presence of a good sagittal diameter of the canal is unlikely to be of importance. In this respect, however one must note the possibility of a narrowed sagittal diameter produced by either hypertrophy or folding of the ligamentum flavum. Huizinga (1952) carried out a biometric survey of 51 lumbar vertebral canals (obtained from the exhumed skeletons of nineteenth century subjects) and recorded the least and the greatest sagittal diameters that could be considered to be normal at the various levels (see Table). A wide variation is possible but the canal is normally narrowest at the level of L 4. Verbiest (1955) measured the sagittal diameter of the vertebral canal at

TABLE

Vertebral level	Sagittal diameter (millimetres)		Variation
	Least	Greatest	
L 1	14	22	8
L 2	13	20	7
L 3	12	27	15
L 4	11	22	11
L 5	12	22	10

operation and concluded that when the diameter is narrow in relation to the above Table even a small disc protrusion may be significant.

One of the major limitations of myelography is in demonstrating postero lateral protrusions. If the transverse diameter of the theca is less than 16 millimetres the chances of demonstrating a postero lateral protrusion are small (Arbuckle, Shelden and Pudenz 1945) but even with a good transverse diameter

and without asymmetry of filling of the root sheaths postero-lateral protrusions extending into the foramina may be present

In the thoracic spine the opaque column spreads out rapidly and disintegrates. This tends to prevent visualization of anything but the grosser lesions. Slowing down the transit of the opaque medium by examining the patient in the supine



FIG 157—Cauda equina showing multiple small metastases from small (and clinically undetected) bronchial neoplasm. Myelogram negative (B) courtesy of Dr F M Peters)

The use of water soluble contrast medium is attractive because by this method the components of the cauda equina can be visualized throughout their length. The method requires spinal anaesthesia. Not all types of contrast media can be used and fatalities have been reported from the accidental use of the wrong material. The complication rate is high, and the opaque medium must be confined to the lumbar region. For these reasons it has not gained wide acceptance. The only medium Myodil (Pantopaque) is virtually non toxic, it is gradually if slowly absorbed and although for medico legal reasons it should ideally be withdrawn in many centres this is not done. Indeed, unless the needle is left *in situ* during the examination, the additional puncture may itself produce complications, as in the case reported by Keats (1956) in which the medium was seen to disappear rapidly and form emboli in the lungs. Violent and uncontrollable coughing occurred but there were no severe symptoms and recovery was complete. To leave the opaque medium *in situ* has the advantage of allowing re examination at intervals and assessment of the results of operation. The method has limitations (Reid and Tutton 1951, McCarthy and Lane 1955) both in the diagnosis of disc protrusion and of other space occupying lesions in the spinal canal.

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(a)



(b)



(c)

FIG 158—Osteomyelitis. Pain in back 5 weeks after staphylococcal chest infection (a) 3 months after onset. Maximum destruction (b) 9 months. Healing process begins (c) 16 months. Considerable regeneration.

position does not assist, because the medium then lies at a distance from the posterior surface of the vertebral bodies

In the cervical spine, transverse bars are very common and their significance in relation to the patient's symptoms is a matter for clinical decision

True localized disc protrusions are fairly easily demonstrated, but the interpretation of asymmetry of the root sheaths when postero lateral osteophytosis is present is difficult. With space occupying lesions of the canal, the classical myelographic appearances associated with the various types and situations of lesions may be simulated by other conditions. Lateral radiography in the supine and prone positions is often of assistance, but even after careful examination and the study of films taken in all projections an erroneous interpretation of the site of a lesion in relation to the cord or meninges may be made. A completely normal myelogram may be obtained in a patient with undoubted clinical signs of root involvement and in such conditions the possibility of multiple small secondary deposits on the cauda equina should be borne in mind (Fig 157)

INFECTIVE LESIONS

Tuberculosis

This subject is considered in Chapter 6. Radiologically two problems are met with. The first is early diagnosis and the second is the estimation of progress in the established case. In Great Britain any reduction in disc space if it is associated with a loss of definition of the vertebral disc surface must invite the suspicion of tuberculosis even if this loss of definition is associated with a slight increase in density. Any progressive loss of disc space in a young person or any suspicion of a localized bulging of the paravertebral shadows in the thoracic spine or generalized bulging of psoas shadow in the lumbar spine must also be closely observed and where necessary further investigated. Oblique films may assist in demonstrating small cortical or marginal lesions and very small areas of destruction in the vertebral disc surface may be shown by tomography. The estimation of progress is largely a matter of meticulously comparing the radiographic appearances in each portion of the affected segment of the vertebral column but care should be taken to ensure that the vertebral bodies above and below for a considerable distance are well shown at each examination in order to check that no spread beneath the anterior longitudinal ligament is occurring.

Pyogenic infections

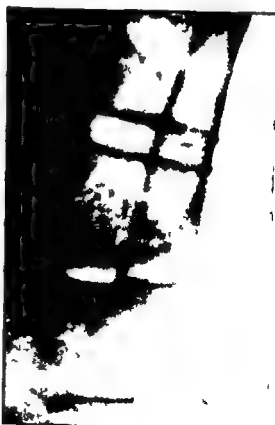
Osteomyelitis in the spine is relatively uncommon and unlike osteomyelitis in long bones, it appears to be less common in childhood than in adult life. The condition may be subacute (Gurr 1946) and may warrant the description 'non suppurative' as in the case of a child reported by Kadd (1951) in whom a staphylococcal lesion appeared in a transverse process after injury. Before the introduction of antibiotics the outlook was poor. In general there is disc destruction with rarefaction of the adjacent vertebral body followed by partial collapse, areas of sclerosis, new bone formation and eventually synostosis. The paravertebral abscess is small compared with that of tuberculosis. Exceptions naturally occur and in some cases a striking regeneration of the affected vertebra is seen (Fig 158). As in all cases of vertebral infection from whatever cause the possibility



(a)



(b)



(c)

FIG 158—Osteomyelitis. Pain in back 5 weeks after staphylococcal chest infection (a) 3 months after onset. Maximum destruction (b) 9 months. Healing process begins (c) 16 months. Considerable regeneration.

of epidural spread must be borne in mind and this subject has been reviewed and the myelographic findings described by Campbell and Silver (1954). The most common aetiological factor would appear to be infection of the urinary tract with or without surgery (Wood, 1954, Liming and Young 1956, Henson and Coventry 1956).

Typhoid and brucellosis

The picture is usually one of multiple disc lesions associated with irregularity and rarefaction of the adjacent body followed by sclerosis and healing with either bony bridging or massive osteophytosis.

Syphilis

Syphilis may affect the spine either directly in the form of a gummatous lesion or indirectly through a neuropathy.

Gummatous lesions are uncommon and the literature has been reviewed by Epstein (1955). There is usually a widespread patchy bone destruction and the response to anti syphilitic treatment is conclusive, the lesion healing by fusion and ankylosis.

The neuropathic changes associated with tabes consist of an irregular sclerosis of the affected vertebra, usually in the lumbar spine, an ill defined cortical margin

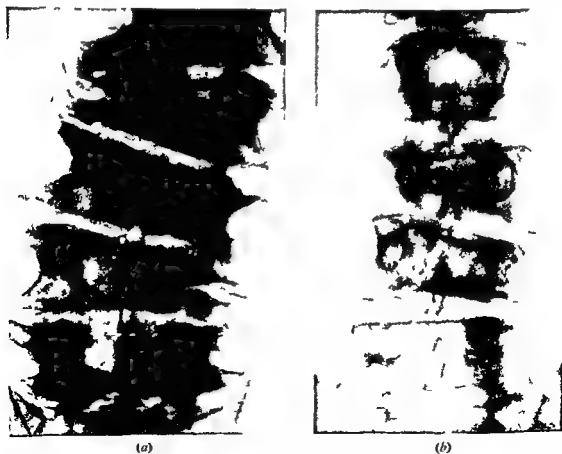


FIG 159—(a) Charcot spine (b) Lymphadenoma

to the body, disc generations and a peculiar toppling appearance due to subluxations of the vertebral bodies in an irregular manner. Occasionally fragmentation of the margins of the bodies occurs.

Parasitic infection

Only hydatid disease is of importance in relation to bone involvement. The condition is not as uncommon as might be supposed. Kienboch (1933) reviewed 86 cases of bone hydatid disease. In these there was involvement of the pelvis in 33, the sacrum in 7, and the vertebral column in 16. He described four main types, primarily in relation to long bone infections, but applicable to the changes found in the vertebral column: (a) an infiltrating lesion producing cortical destruction without reaction, (b) an expanding lesion with a cortical outline and bony trabeculae producing a cystic appearance, (c) a fibrous sac radiologically only a soft tissue mass, and (d) a subperiosteal type producing a cortical destruction with a large lateral outgrowth which may or may not have a calcified periphery. In the spine involvement of several vertebrae and the adjacent ribs is common, but on the whole the intervertebral discs tend to be spared. However, in some cases there is complete destruction over a considerable portion of the column and the question as to whether or not the discs are spared is entirely academic. Latham (1953) reported a case of hydatid disease in the spinal canal with a complete block on myelography.

Fungus infections of the spine have been recorded on many occasions and include cases of actinomycosis, blastomycosis, coccidiomycosis and torulosis. In actinomycosis the affected vertebrae have a honeycomb appearance and eventually marked bony sclerosis may be present surrounding the infected areas. The whole of the vertebral column may be involved. The intervertebral discs are as a rule spared and collapse is not common (Lubert, 1944). In the other fungus infections, the general rule is that of a lytic lesion with paravertebral abscesses and lesions in the spinal canal.

ACROMEGALY

Deposition of new bone around the circumference of the vertebral body occurs in some cases of acromegaly, particularly in the lower thoracic and upper lumbar areas. It is also seen in long standing osteoarthritis and in osteochondritis where anterior discal rupture has occurred.

RADIATION HAZARDS

The problems of radiation damage to the individual and to succeeding generations have recently received wide publicity.

Most children born since World War II will be examined radiologically at some time during their life even if only as a result of exhortations to attend mass radiographic centres for chest radiographs (where they will receive 5-16 times the dose that they would have received during a conventional radiological examination). The major problem, however, is that of radiography of the spine, the pelvis and the hips. In these cases the doses received by the gonads are high and, for all practical purposes in Great Britain at any rate, no attempt has commonly been

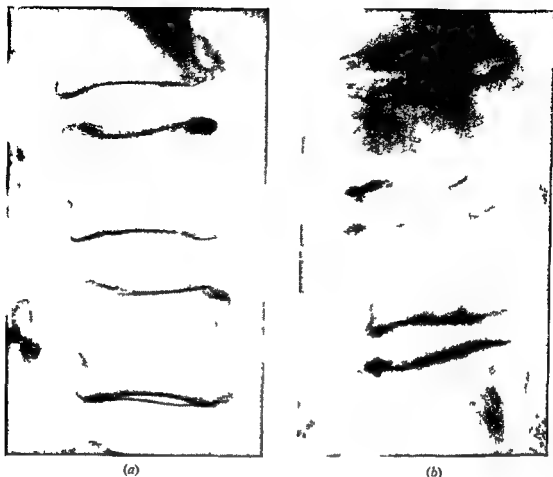


FIG 160—Deposition of new bone (a) Osteoarthritis (b) Acromegaly

made to reduce the dose to the ovaries or the testes by the use of lead protection. The possibility no matter how remote of late neoplastic change in the skin following radiography of the lumbar spine must be borne in mind. So far as the writer is aware no such lesion has been observed. Such an unfortunate occurrence would be a matter of chance and the odds against its happening will decrease as the number at risk increases.

A few figures may be of interest: 3 000 r administered to one area of the skin over a period of two weeks will produce blistering. 350-500 r given in a single dose will produce epilation. A lateral film of the lumbo sacral angle will deliver to the skin from 8 to 35 r depending on the size of the patient, the apparatus and the technique used. In a difficult patient three attempts may be made to obtain a satisfactory film. If followed by lateral tomography and a re check after 14 days the dose may well be between 88 and 365 r. This does not include the supplementary doses received from the antero posterior films of the lumbar spine and pelvis, the oblique films of the sacro iliac joints and the full lateral film of the lumbar spine. Oddly enough the higher doses will be received in those departments attempting to produce the best work using high ratio grids, high definition intensifying screens and standard films. High kilovolt units (up to 120 kilovolts) produce a satisfactory reduction in the skin dose down to one third or

one quarter but reverting to gonad doses they offer little improvement and indeed may be a disadvantage since the total radiation absorbed in the body is of the same order and the scattered radiation available to reach the gonads from a distance is greater.

The increased incidence of leukaemia in patients with ankylosing spondylitis treated by x rays is well known. Radiography during pregnancy may or may not be the cause of some congenital abnormalities certainly therapeutic doses have produced abnormalities. Recent surveys by Stewart Webb and Hewitt (1958) seem to suggest that diagnostic irradiation of the pregnant abdomen increases the incidence of malignant disease in the children during their first 10 years of life but that this alone is not responsible for the general increase in childhood malignancy. At present it is difficult to say with any certainty



FIG 161—Tomograph of radio-necrotic collapse of two lobes after radiotherapy for new growth of the bronchus. This patient returned to work as a window cleaner with these lesions which were almost symptomless.

that any particular diagnostic procedure is dangerous to the individual. We do know that where the skin has received a dose sufficient to produce blistering and subsequent pigmentation if the patient lives long enough he is at some risk to the development of neoplasm in that area. The dosages received by some patients from diagnostic radiation is tending to extend into the therapeutic levels. It would be unwise to wait for time to prove or disprove the dangers and in general therefore the most careful consideration of methods of reducing radiographic dosage both to the individual and to the population should be made.

With therapeutic dosages (1 000-6 000 r depending on the time during which the dose has been administered) bone changes are sometimes encountered. In

the young dosage above 1,000 r is likely to alter bone growth in the primary ossific centres in vertebral bodies, and at the epiphyses, with consequent wedging in the spine and shortening and deformity in the limbs

At higher levels in the adult radiation necrosis is not uncommon. In general one associates radiation necrosis with severe pain and sequestration but this is usually due to infection in dead tissue and occurs mainly in the upper air passages in the mouth and in superficial bone and cartilage. In the deeper situations for example the femoral neck and pelvis (uterine neoplasms) and the thoracic spine (bronchial and oesophageal neoplasms) there may be no symptoms or pain of varying severity for months or years. With fracture, pain usually occurs to a greater or less degree. This possibility must be remembered in the case of the spine as the collapse is indistinguishable, apart from its progress, from that due to a metastasis. At 6 000 r and above the possibility of necrosis of the spinal cord arises and localized lesions have been recorded following such dosages in the cervical spine by Boden (1948).

SUMMARY

A clinical diagnosis is made on the results of many investigations, most of them important and essential. In spinal disabilities radiography is one such investigation. It cannot be omitted, but it is not the all important diagnostic tool which it was previously thought to be. The great majority of abnormalities which it finds are aging phenomena and it has become increasingly obvious that the degree of disability and the bony appearances do not always correspond. The radiologist describes and can make limited deductions from the appearances of the vertebral column. The clinician however, must make a diagnosis based on the patient's symptoms and the findings of physical and radiological examinations. Faced with the volume of work which now passes through his department the radiologist's most difficult task is the maintenance of good standards in technique and scrutiny. Given a specific problem any radiological team can produce excellent films and a competent differential diagnosis. But the objective should always be the detection of those lesions in which an early diagnosis may be important. The limitations of plain radiography have been commented upon. Nevertheless its main value in practice is the exclusion of infective and neoplastic conditions and it is an essential preliminary to any manipulative procedure.

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CHAPTER 12

MANAGEMENT OF SPINAL CORD INJURIES

L. GUTTMANN

STATISTICAL ANALYSIS

Material

THE TOTAL number of paraplegics treated at the Stoke Mandeville National Spinal Injuries Centre during the period February, 1944 to September 1956 is shown in Table I, from which it is clear that the great majority of cases of paraplegia are of traumatic origin. Included under the heading 'Miscellaneous' are those patients whose paraplegia was caused by tumour, vascular processes, congenital mal-developments (for example spina bifida), tuberculosis, disseminated sclerosis and so on.

TABLE I

NATIONAL SPINAL INJURIES CENTRE, STOKES MANDEVILLE HOSPITAL
PARAPLEGICS TREATED FEBRUARY 1944 TO SEPTEMBER 1956

	Injuries	Transverse myelitis	Poliomyelitis	Miscellaneous	Total
<i>Cervical</i>	104	12	41	46	203 (14%)
Complete	31	1	—	4	36
Incomplete	73	11	41	42	167
<i>Thoracic 1-5</i>	107	12	1	31	151 (10%)
Complete	82	5	—	16	103
Incomplete	25	7	1	15	48
<i>Thoracic 6-12</i>	514	64	57	145	780 (52%)
Complete	405	38	14	56	513
Incomplete	109	26	43	89	267
<i>Cauda equina</i>	322	6	1	37	366 (24%)
L1-5	275	4	1	30	310
S1-5	47	2	—	7	56
<i>Total</i>	1047 (70%)	94 (6%)	100 (7%)	259 (17%)	1500 (100%)

Mortality

The dramatic change in the mortality rate of spinal paraplegics now compared with the past is shown in Table II.

These statistics include all those who died either at Stoke Mandeville or after discharge from the Centre to their own homes or other institutions. The uncorrected figures also include those who died not as a result of their spinal injury and its complications (embolus following thrombosis or sepsis from urinary infection and pressure sores for example) but from other causes such as cancer, tuberculosis, road accidents, cerebral haemorrhage, coronary thrombosis and so on. It is of particular interest that the death rate of traumatic paraplegics from World War II still continues to be very low. If the corrected figures are considered, it will be seen that the majority of paraplegics from this war have already survived for as long as 13 to 18 years. Moreover, it must be remembered that many of the war wounded arrived at the Centre in a deplorable

TABLE II

NATIONAL SPINAL INJURIES CENTRE STOKE MANDEVILLE HOSPITAL
MORTALITY FEBRUARY 1944 TO SEPTEMBER, 1956

A Total Material—1 500 Cases			
150 Deaths out of 1 500			10%
116 (corrected figure)*			7 7 /
II Injuries—1 047 Cases			
110 Deaths out of 1 047			10 5 /
86 (corrected figure)			8 2 /
<i>Service Cases and Pensioners</i>		<i>Civilians</i>	
84 Deaths out of 621	13 5 /	26 Deaths out of 426	6 1 %
68 (corrected figure)	10 9 %	18 (corrected figure)	4 2 %
<i>Analysis of traumatic Service cases and pensioners</i>			
(1) World War I			
10 Deaths out of 51			19 6 %
6 (corrected figure)			11 8 %
(2) World War II			
73 Deaths out of 465			15 7 %
61 (corrected figure)			13 1 %
(3) After war period			
1 Death out of 105			0 95 /

After deducting those cases where death was due to causes other than spinal paraplegia

condition with extreme malnutrition huge bedsores gaping wounds infected bladders and kidneys profound flexor spasms contractures of tendons and joints and thoroughly demoralized. If the mortality rate of the victims of World War II is compared with the extremely low rate of the post war period it will be seen how new methods of treatment have altered the outlook.

Because of the high survival rate the problem of paraplegia today can no longer be considered merely as a medical one. It has become a social problem of increasing importance from year to year. The utmost effort on the part of everyone concerned with the management of paraplegics at all stages is required to return as many paraplegics as possible to useful work and employment.

Domestic and industrial resettlement

The figures for domestic and industrial resettlement are very encouraging indeed. It may be noted that many of the full time employed paraplegics are working in factories side by side with able bodied people. Others are employed in business the professions clerical work farming as craftsmen and in many other occupations. Paraplegic women can carry out housework and look after their families from their wheelchairs and some of the paraplegic children can receive education in ordinary schools. As far as the permanent disability of spinal paraplegia is concerned only a very small percentage of paraplegics—that is those with high lesions involving paralysis of both arms in addition to the legs and trunk—can be considered unemployable.

From all the data given it is obvious that it is not the Spinal Unit team only that is responsible for the rehabilitation of paraplegics but that the medical practitioner also has an increasingly important part to play in the aftercare of paraplegics once the patients have been discharged to their own homes or to special settlements. This

INITIAL MANAGEMENT

also applies to the representatives of social services local housing committees and in particular the Ministry of Labour. And last but by no means least it applies to the public generally and to employers in particular. With regard to the latter there is still a good deal of education necessary for many employers if they see a man in a wheelchair tend to assess only what the man has lost and not what he still has remaining. The author's experience has been that once the first obstacle of finding employment for a paraplegic has been overcome his employer will find that he can hold down his job with any able bodied comrade.

Absenteeism of paraplegics from work is usually no greater than that of able bodied workers. Very few have had to give up work because of complications of spinal paraplegia. It is distinct from pre existing problems of personality.

INITIAL MANAGEMENT

FIRST AID

In all discussions in the past on first aid in spinal injuries the need for standardized simple instruction has been readily recognized and, from all the experience gained the following conclusions can be drawn

(1) A patient with a suspected spinal fracture following a road or industrial accident should be warned not to move.

(2) At least 3 and if possible 5 people should move the patient from the place of accident. It is of utmost importance that all movements by the first aid party be carried out slowly and with the greatest care not to bend the patient either backwards or forwards. Moreover meticulous care should be taken that all movements are carried out simultaneously so that the patient is turned or lifted in one piece. This principle of moving the patient in one piece should be strictly adhered to whether the lifting be done by hand or by straps placed underneath the patient.

(3) If the injured person is found in any position other than on his back he should be gently turned on to his back in one piece and a cushion or improvised soft support large enough to maintain the normal contour of the spine placed under the small of his back. All patients with spinal injuries except those who are unconscious can be safely and comfortably transported in the supine position. In suspected injuries of the upper thoracic and cervical cord this is absolutely vital because of the involvement of the respiratory apparatus. In cases with a cervical injury the person who is holding the head must apply slight traction while taking care that the head is maintained slightly extended and not bent forwards. There is no need to transport an individual with a spinal fracture of the thoraco lumbar region in the prone position. In fact as a spinal lesion is not infrequently associated with other fractures or with collapse of one side of the lung the prone position may even be harmful. The only time a patient with a suspected spinal injury should be transported in any position other than supine is if he is unconscious when in order to avoid aspiration of saliva it is safer to transport him in a lateral position.

(4) The patient with a spinal injury should be transported on a rigid stretcher. A pillow or other soft support should be placed underneath the small of his back to produce slight hyperextension of the spine. Hard objects should be removed from the patient's pockets and areas of prominent bone should be padded. The simplest way to protect the heel and the head of the fibula from pressure is to place pillows or a blanket underneath the calves. For transportation the feet and legs should be bound together at the ankles and thighs. The patient should be covered with blankets, to

preserve body heat and diminish general shock. In no circumstances should hot water bottles be placed on any part of an injured person with paralysis.

(5) Morphine should not be given indiscriminately and is indicated only in the presence of severe root pain. Its administration in cervical lesions and other cases where there is a suspicion of lung collapse may prove harmful if not disastrous.

(6) Before transferring the injured person to a spinal unit or the nearest hospital the first medical attendant of a traumatic paraplegic should ascertain as soon as possible the level of the lesion and in particular the completeness of the motor and sensory paralysis. A written statement about his observations should accompany the patient.

HOSPITAL TREATMENT

Management of the fractured spine

The guiding principles in the initial management of traumatic paraplegics are the greatest gentleness and an avoidance of hasty surgical and manipulative procedures.

As a rule, the person with a spinal injury is taken to the nearest hospital for treatment of shock and initial management of the broken spine. The hospital should get in touch immediately with the nearest spinal unit with a view to obtaining specialized advice as to the immediate steps to be taken and arranging for an early transfer to such a unit where there undoubtedly exist the best facilities for carrying out the whole rehabilitation of the paralysed.

When the patient has arrived at the hospital the principle of care in lifting and turning the patient *in one piece* by several attendants from the stretcher on to his bed, x-ray or operating table must be preserved as it is the main factor in preventing further damage.

Postural reduction on pillow or sorbo packs and regular turnings

The routine procedure at Stoke Mandeville Spinal Centre in dealing with traumatic paraplegia in the initial stage after fracture or fracture dislocation is the employment of postural reduction on sorbo packs and regular (two hourly) turnings the technique of which is as follows:

The patient is placed on sorbo packs with 1, 2 or 3 additional pillows underneath the fracture to produce hyperextension of the spine in the physiological position to restore as far as possible the normal curvature of the spine (Fig 162). From the



Fig 162—Initial treatment of traumatic paraplegia. The patient is nursed on sorbo packs with additional pillows under the fracture to produce hyperextension of the spine.

INITIAL MANAGEMENT

basic supine position the patient is turned first on to one side the hyperextended position being maintained by the use of a sandbag to support the pillow in the back then back to the supine position and then on to the other side every 2 hours day and night (Fig 163). The turning is carried out by 3 or 4 attendants working under the guidance of the medical officer sister or nurse in charge of the case and they are made fully aware of the details of the fracture and the importance of carrying out the lifting and all movements simultaneously so that the patient is turned in one piece without dragging.



FIG 163—The hyperextended position being maintained with the patient on his side by the use of a sandbag to support the pillow in the back.

With this method of postural immobilization and regular turning ordinary compression fractures of the spine can be kept safely in position and fracture dislocations can be brought into satisfactory alignment and stabilized in a period of between 6 and 10 weeks. It may be emphasized that even unstable fractures can be reduced and kept in position by this method (Guttmann 1954 1956). Fig 164 shows the result of postural reduction with immobilization and regular turning in a case of fracture dislocation of the third lumbar vertebra resulting in a severe cauda equina lesion. The patient made an excellent recovery and on discharge from hospital was able to walk without the aid of sticks.

It may be noted that this method of postural reduction has proved very satisfactory not only in fractures and fracture dislocations of the thoraco lumbar spine but also of the cervical spine. Before head traction is applied by Crutchfield tongs an attempt should be made to reduce the fracture by placing a small hard pillow under the nape of the neck fixing the head with sandbags on either side. The patient can then be turned in the way described above. The person holding the head during the turning must be an experienced nurse for she must ensure that the head and trunk are turned from the supine to the lateral position in one piece. This simple method has proved successful in reducing moderate dislocations of cervical vertebrae (Fig 165). However if this method is unsuccessful head traction with Crutchfield tongs has been employed as the method of choice in this Centre with good results.

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FIG 162—Initial treatment of traumatic paraplegia The patient is nursed on sorbo packs with additional pillows under the fracture to produce hyperextension of the spine

INITIAL MANAGEMENT

In the supine position the patient is turned first on to one side the hyperextended position being maintained by the use of a sandbag to support the pillow in the back then back to the supine position and then on to the other side every 2 hours day and night (Fig 163). The turning is carried out by 3 or 4 attendants working under the guidance of the medical officer sister or nurse in charge of the case and they are made fully aware of the details of the fracture and the importance of carrying out the lifting and all movements simultaneously so that the patient is turned in one piece without dragging.



FIG 163—The hyperextended position being maintained with the patient on his side by the use of a sandbag to support the pillow in the back.

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8 12 54



23 12 54



22 2 55



6 6 55

FIG. 164 — Fracture dislocation of the third lumbar vertebra. Here is seen the result of postural reduction with immobilization and regular turning.

With this method of immobilization of paraplegics on pillow or sorbo packs and regular turning every 2 hours day and night the development of pressure sores has become unknown and in cases admitted with sores already developed healing has taken place in a very short time. Moreover in patients with fracture dislocations of the spine which were not reducible and which showed an incomplete lesion of the cord or cauda equina postural immobilization on pillow packs



11 1 57



8 2 57

FIG 165—Postural reduction used in a case of dislocated cervical vertebra



25 4 57

with regular turning has proved satisfactory in ensuring progressive recovery of the cord or roots of the cauda equina even in the presence of considerable bony displacement. This applies not only to the cervical and lumbar spines but also to the mid thoracic spine, where normally the vertebral canal is much narrower. There is, thus, no reason for any surgical or manipulative measures in the initial stage of treatment of traumatic paraplegia. The great advantage of postural

reduction in the initial treatment is that it also ensures a speedy recovery of the back muscles from the direct damage they sustained at the time of injury. This is of utmost importance for restoring and maintaining the stability of the spine.

The Stryker turning frame

The Stryker frame and similar types of turning bed (for example, the Emesay) have been introduced to facilitate turning of paralysed patients.

However, their main disadvantage is that they are very narrow and allow turning from the supine to the prone position only. Therefore, they are unsuitable for the immediate or early management of the fractured spine, especially fracture dislocations of the lower thoracic and lumbar vertebrae, as the hyperextension of the broken spine achieved by postural reduction in the supine position with the aid of a pillow or sorbo support cannot be maintained once the patient is turned into the prone position because this flattens out the lumbar curvature and prevents stabilization of the broken vertebrae.

External fixation and immobilization by plaster cast or plaster bed

External fixation of the spine in a plaster cast in cases of traumatic paraplegia following fractures is, as a rule, unnecessary as an immediate measure.

Only in selected cases of unstable fracture dislocation, where displacement recurs after postural reduction and immobilization or for transport of a patient from the first aid post or hospital to a spinal unit, may this form of external fixation be considered.

Plaster beds, which were advocated during World War II for the initial treatment of traumatic paraplegia following fractures and for the prevention of pressure sores, have proved utterly unsatisfactory in the author's and other workers' experience and this method is to be condemned for the initial treatment of traumatic paraplegia.

Surgical reduction and fixation by bone graft or metal plating

At the beginning of World War II, operative reduction in certain forms of fracture dislocation was recommended—in particular by Albee (1940), using bone grafts and by J. Taylor (1941). Open reduction and fixation of the broken vertebra by metal plating has been revived by Holdsworth and Hardy (1953) who condemned manipulative reduction as being usually unsafe and advocated internal fixation in paraplegics with fracture dislocation of the thoraco-lumbar spine. They advocated bolting 2 metal plates over 1 or more spinous processes above and below the level of the dislocation. They described this method as simple, safe and effective. Pennybacker (1953) suggested as likely that this kind of internal fixation would prevent progressive angulation and other deformities when the upright posture and weight bearing were resumed. Dick (1953) stated that the spine can be stabilized in this way so securely that ordinary nursing handling is absolutely safe and there is no danger of further damage to recovering nerve roots.

Since this method was recommended, 41 cases on whom open reduction with metal plating had been carried out as initial treatment have been admitted to the National Spinal Injuries Centre and it may be noted that this operation had in the great majority of cases been performed by expert surgeons. This considerable material enables certain conclusions to be drawn in the light of the claims made by the advocates of this method.

Simplicity of method—On anatomical grounds it is apparent that plating may be difficult if one or several spinous processes are broken and that secure fixation

INITIAL MANAGEMENT

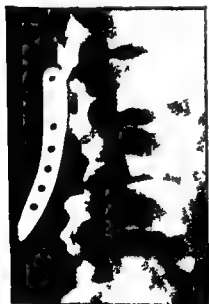
must involve the use of plates extending over at least 4 vertebrae with the associated interference with the back muscles. Holdsworth and Hardy themselves admit difficulties and point out that the surgeon has to exercise ingenuity in fixing the plates to the lamina.



10 7 53



16 7 53



10 9 53



23 11 53

FIG 166—Internal fixation by metal plates which did not prevent re dislocation. The plates later had to be removed.

Prevention of re dislocation—Fig 166 shows in a case of fracture dislocation that while the reduction of the dislocated vertebra was at first perfect re dislocation

reduction in the initial treatment is that it also ensures a speedy recovery of the back muscles from the direct damage they sustained at the time of injury. This is of utmost importance for restoring and maintaining the stability of the spine.

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Since this method was recommended, 41 cases on whom open reduction with metal plating had been carried out as initial treatment have been admitted to the National Spinal Injuries Centre and it may be noted that this operation had in the great majority of cases been performed by expert surgeons. This considerable material enables certain conclusions to be drawn in the light of the claims made by the advocates of this method.

Simplicity of method—On anatomical grounds it is apparent that plating may be difficult if one or several spinous processes are broken and that secure fixation

stages and it was thought that this might be prevented by metal plating. That this is not the case is shown by Fig. 168.

Safety of method—These few examples may suffice to disprove the assumption that internal fixation by plating wire or bone grafts is a safe method for the stabilization of the fractured spine particularly if ordinary nursing handling, as emphasized by Dick, is employed. The author considers it very hazardous to dispense after internal fixation, with that meticulous care which is so imperative in the management of a paraplegic patient.

Efficiency of the method in promoting recovery of spinal roots and in preventing further damage—From the literature as well as from our own observations following open reduction and plating there is no proof that this method is in any way superior to other methods of reduction and immobilization, and it may be noted that there were cases in our material where it was quite evident from the previous

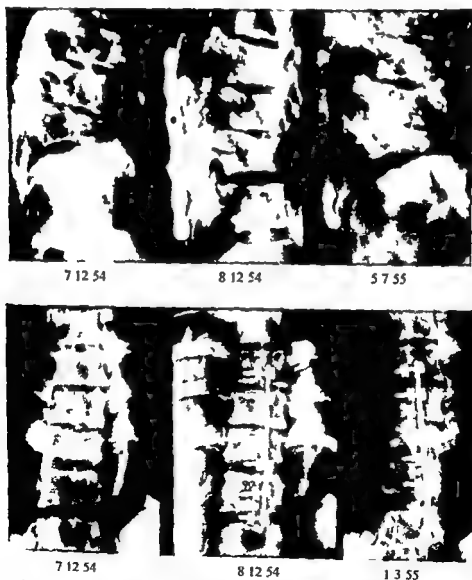


FIG. 168—Collapse of erected vertebra in a case of compression fracture not prevented by metal plating.

was not prevented by the internal fixation. In fact the plates had to be removed after the patient's admission to the Centre, as they had been driven backwards by the re-dislocated vertebra and were not only producing discomfort and pain, but were in danger of breaking through the skin.



25 9 53



5 10 53



19 10 53



30 10 53

FIG 167—Internal fixation by metal plates. In this case even the use of very long plates failed to prevent progressive angulation.



21 5 54

Prevention of progressive angulation—It will be readily understood from a purely mechanical point of view that if the broken vertebra is fixed to its neighbours by a wire or short plates only progressive angulation cannot be prevented. But even in cases with very long plates progressive angulation in later stages is very conspicuous (Fig 167).

Prevention of collapse of the erected broken vertebra—In compression fractures in which the compressed vertebra was erected by conservative methods such as plaster or postural reduction collapse of the erected vertebra may recur in later

the neurological symptoms of these cases treated by laminectomy. On the contrary they emphasize that aggravation of symptoms was extremely frequent.

Immediate mobilization without reduction

The author however does not support that school of thought which ignores completely the fractured spine resulting in spinal paraplegia. Sutcliffe Kerr (1936) advocated in complete transverse spinal lesions that the paraplegic be sat up within a few days after injury. His views are based on the conception generally held that if within 48 hours after injury the lesion is still complete there is no hope of any recovery. Experience has shown that this is not always so as recovery of function has been known to occur even after several weeks. Therefore the author considers as hazardous this swing of the pendulum to the extreme conservative approach which neglects the vertebral deformity and allows the patient to be sat up a few days after injury. As far as possible alignment of the broken spine should be attempted and this can be achieved in many cases by the gentle method of postural reduction.

Treatment of shock

The combating of general shock as an immediate measure is of particular importance in traumatic paraplegia as the restoration of a good blood pressure will help to counteract the lowering of tissue resistance to pressure caused by interruption of the spinal centres and pathways subserving vasomotor control in the paralysed parts of the body. The easiest and quickest way of combating shock is a full blood transfusion. The sooner the shock is overcome the sooner is it possible to transfer the patient to a spinal unit or hospital equipped for the treatment of paraplegia. In recent years traumatic paraplegics have been transferred in increasing numbers to Stoke Mandeville within hours or 1 or 2 days of their injury. This is the ideal procedure.

The application of heat in the treatment of shock in paraplegia has to be considered with the greatest caution particularly where a lesion of the cervical cord is concerned because this renders the patient poikilothermal owing to interruption of the thermoregulatory pathways. In no circumstances should a hot water bottle even with a cover be applied to a paraplegic patient whatever the level of injury.

Management of the paralysed bladder

If the patient's condition does not permit immediate transfer to a spinal unit the management of the paralysed bladder in addition to the prevention of bedsores becomes a most important responsibility pending the patient's transfer.

The immediate management of the paralysed bladder does not include intervention by instrumentation as a rule the bladder is not so distended as to warrant immediate drainage by any method. Moreover in many cases of spinal injury the rate of renal secretion is retarded. In particular there is no indication for performing immediate suprapubic drainage. It has its use only in those patients in whom the urethra has sustained a direct injury at the time of accident.

During the early period repeated attempts should be made to overcome retention and to elicit voiding by gentle manual pressure upon the bladder region combined with digital massage per rectum. The fear that manual pressure may rupture the bladder has proved to be exaggerated.

medical notes that the clinical symptoms actually increased in intensity following surgical intervention. One of the great disadvantages of this method is that it produces rigidity of the spine which is contrary to one of the main principles of rehabilitation—mobility. Moreover in a relatively large number of patients this procedure produced constant pain which increased the immobility. In our total number of 41 platings 15 plates had to be removed by us the main indication for this being unbearable pain.

Conclusion—From all these observations the conclusion is evident that the method of open reduction followed by internal fixation by metal plating does not represent a satisfactory initial treatment in stabilizing the fractured spine with cord or cauda equina involvement. Not only is this method quite unnecessary in the initial treatment of the vast majority of fracture dislocations but it actually has disadvantages compared with the closed method of postural reduction followed by careful immobilization and regular turning. The open reduction by internal fixation with plating or bone grafts has its indication only in very carefully selected cases—that is in the most severe types of fracture dislocation especially those with profound lateral displacement described in a case by Merle d Aubigne Benassy and Ramadier (1956).

Laminectomy

In agreement with other workers with extensive experience in traumatic paraplegia the writer is strongly opposed to laminectomy as an immediate measure in spinal injuries.

Laminectomy represents an additional trauma of considerable degree to the paraplegic patient and no doubt prolongs the period of shock and loss of vasomotor control. It increases the risk of thrombosis and embolus especially in cases with associated injuries to the muscles or skeletal structures in other parts of the body. Furthermore it increases the instability of the spine by removing the posterior arch of the vertebra and by further damaging the back muscles at the site of the injury.

The presence of a subarachnoid block in the early stages following spinal injury is of no diagnostic value in differentiating between oedema haematoma or pressure from the dislocated bone nor in the narrowing of the spinal canal as shown in the x rays a proof that the neurological symptoms are merely the result of pressure. In fact the experience of many authors has been that decompression laminectomies are of no avail. Moreover there is abundant evidence that an ill considered laminectomy may transform an incomplete lesion of the spinal cord into a complete one.

The view of Covalt Cooper Hoen and Rusk (1953) that laminectomy carries a low mortality and morbidity rate can hardly be accepted as a sound argument in favour of routine laminectomies in the early stages of traumatic paraplegia as suggested by these authors. Another argument of theirs in favour of laminectomy is that unless it has been determined by surgical exploration that the spinal cord has been transected one cannot conclude during the early weeks after injury that a patient is permanently or irreversibly paraplegic. It is questionable whether it is justified to submit a paraplegic in the early stages to an exploratory operation of this nature having regard to the fact that no conclusions can be drawn at this juncture even if the spinal cord is found to be intact.

Merle d Aubigne Benassy and Ramadier (1956) have confirmed the author's conservative view against routine laminectomies in traumatic paraplegia. In their experience of 56 cases of traumatic paraplegia they have never observed the slightest improvement in

the neurological symptoms of the 6 cases treated by laminectomy. On the contrary they emphasize that preservation of symptoms was extremely frequent.

Immediate mobilization without reduction

The author however does not support that school of thought which ignores completely the fractured spine resulting in spinal paraplegia. Sutcliffe Kerr (1956) advocated in complete transverse spinal lesions that the paraplegic be sat up within a few days after injury. His views are based on the conception generally held that if within 48 hours after injury the lesion is still complete there is no hope of any recovery. Experience has shown that this is not always so. Recovery of function has been known to occur even after several weeks. Therefore the author considers as hazardous this swing of the pendulum to the extreme conservative approach which neglects the vertebral deformity and allows the patient to be sat up a few days after injury. As far as possible alignment of the broken spine should be attempted and this can be achieved in many cases by the gentle method of postural reduction.

Treatment of shock

The combating of general shock is an immediate measure is of particular importance in traumatic paraplegia as the restoration of a good blood pressure will help to counteract the lowering of tissue resistance to pressure caused by interruption of the spinal centres and pathways subserving vasomotor control in the paralysed parts of the body. The easiest and quickest way of combating shock is a full blood transfusion. The sooner the shock is overcome the sooner is it possible to transfer the patient to a spinal unit or hospital equipped for the treatment of paraplegia. In recent years traumatic paraplegics have been transferred in increasing numbers to Stoke Mandeville within hours or 1 or 2 days of their injury. This is the ideal procedure.

The application of heat in the treatment of shock in paraplegia has to be considered with the greatest caution particularly where a lesion of the cervical cord is concerned because this renders the patient poikilothermic owing to interruption of the thermoregulatory pathways. In no circumstances should a hot water bottle even with a cover be applied to a paraplegic patient whatever the level of injury.

Management of the paralysed bladder

If the patient's condition does not permit immediate transfer to a spinal unit the management of the paralysed bladder in addition to the prevention of bedsores becomes a most important responsibility pending the patient's transfer.

The immediate management of the paralysed bladder does not include intervention by instrumentation as a rule. The bladder is not so distended as to warrant immediate drainage by any method. Moreover in many cases of spinal injury the rate of renal secretion is retarded. In particular there is no indication for performing immediate suprapubic drainage. It has its use only in those patients in whom the urethra has sustained a direct injury at the time of accident.

During the early period repeated attempts should be made to overcome retention and to elicit voiding by gentle manual pressure upon the bladder region combined with digital massage per rectum. The fear that manual pressure may rupture the bladder has proved to be exaggerated.

If voluntary or reflex function has not developed within 24 to 36 hours of a spinal injury drainage of the bladder is indicated. This is carried out by the no touch technique of urethral catheterization under scrupulous aseptic conditions. At first intermittent catheterization is employed every 8 to 12 hours according to fluid intake and the rate of renal secretion. A Foley catheter is used size 14 to 16 in adults and size 12 to 14 in children (French gauge). The reason for first using intermittent catheterization and not an indwelling catheter is to allow the urethral mucosa to become accustomed gradually to the foreign body. It must be remembered that in the stage of spinal shock and flaccidity all tissues including the urethral mucosa have lost their tone and the threshold of tissue resistance to pressure is lowered. With incomplete lesions in which an early return of bladder function can be expected intermittent catheterization allows distension of the bladder which is a strong stimulus for promoting early return of bladder activity.

From the start the patient is given small doses of sulphatriad and oxytetracycline or chlortetracycline.

If infection occurs this is the time to continue the urethral drainage by an indwelling catheter combined with either bladder or tidal drainage. The best type of indwelling catheter is the self retaining Foley catheter size 14 to 16(FG) with 5 millilitre balloon. It must be emphasized that at first the indwelling catheter must be changed every other day and later on at intervals of 2 or 3 days. In order to free the urethra from deposits it should be washed out either with flavazole 1 in 2 000 or with a solution of chloramphenicol before inserting the new indwelling catheter. Such washouts are especially necessary when urethritis has developed. Once the automatic function of the bladder has returned the indwelling catheter should be withdrawn and intermittent catheterization instituted. This should be continued until detrusor action is powerful enough to empty the bladder completely or leave only a small volume of residual urine. The same principle applies to cauda equina lesions when voluntary micturition becomes possible by pressure on the abdominal wall. Frequent residual urine tests are essential during the early stage of bladder re education. This regime has prevented serious complications in most of our cases.

SPECIAL MEDICAL ASPECTS IN THE LATE TREATMENT AND CARE

Nutrition

Many paraplegics admitted to this Centre several weeks or months after injury arrived with varying degrees of malnutrition owing to depletion of the body's protein reserve which resulted from infection of pressure sores and of the urinary tract. Constant attention must be directed not only to overcoming malnutrition during the early stages of paraplegia but also to maintaining a good state of nutrition after the patient's discharge to his own home. This is of particular importance to those paraplegics who are still suffering from chronic infection of the urinary tract or from renal deficiency with increased protein excretion. On the other hand obesity which has been observed in certain paraplegics in later stages (particularly those who relapsed into inactivity at home and did not carry out regular exercises) must also be avoided and treated by special diets to reduce the risk of hypertension and other vascular disturbances.

Care of bladder and bowels

In later stages of paraplegia once the isolated cord has regained its automatic function or in cauda equina lesions micturition occurs from abdominal straining attention has to be given to the voiding mechanism of the bladder. Regular residual urine tests are carried out to ascertain the amount of residual urine. This should never be allowed to exceed 2 ounces. It is beyond the scope of this chapter to describe in detail surgical procedures such as transurethral resection of the bladder neck, pudendal nerve block or resection or low intrathecal alcohol block which have been developed to combat retention of urine in the later stages of paraplegia. These procedures have proved successful in individual cases but they all have their advantages and disadvantages.

Paraplegics at the National Spinal Injuries Centre are trained in the self care and hygiene of their bladders and certain patients are even trained in self catheterization by the non touch technique. They are taught how to check their residual urine and to remove deposits and control chronic infection of the bladder by regular wash-outs. Patients with incontinence have been taught how to overcome this complication by correlating frequency of micturition with the amount of fluid intake. To be socially acceptable however the majority of paraplegics need a urinal. There are several types recommended.

The most careful attention must be given by the family doctor to any flare up of active urinary infection. This is not necessarily manifested by pyrexial attacks but can be developed slowly and without any rise in temperature. The chronic type of infection however can be recognized by certain symptoms such as loss of appetite, feeling liverish, feeling off colour and either increased irritability or lethargy. These are important symptoms of septic absorption and should always be regarded seriously by the medical attendant. Steps to ascertain the type and degree of infection must be taken at once.

The care of the bowels also needs attention in paraplegics. There is no doubt about the relationship between neglect of the bowels and incidence of urinary infection. On discharge from the Spinal Centre the paraplegic has either attained his own individual habit for opening the bowels or the doctor is informed about the steps to be taken to maintain regular bowel action. Chronic constipation in paraplegics should be avoided.

Vascular disturbances

These may develop in both cord and cauda equina lesions and are caused either by exposing the feet and legs even though covered to an open or electric fire or on the other hand by exposing the insufficiently protected paralysed limbs to cold—especially if the paraplegic keeps his paralysed legs continuously in the same position and fails to exercise them.

The sexual problem

Amongst the psychological effects of traumatic paraplegia the fear that sexual function has been lost is paramount. However experience has shown that the general belief that every paraplegic is impotent and infertile is not correct. Therefore the advice given by many doctors to paraplegics not to marry is often unjustified. Moreover we are in a better position today to ascertain the chances of

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general treatment of the patient with special reference to nutritional deficiency anaemia and general infection and local treatment of the sore the principle of which is that a sore should be treated as an open septic wound, therefore the sloughs have to be removed. The local infection is treated according to the type of bacteria and today the various antibiotics which are used in solution and not as ointments are at our disposal. Chemicals are avoided in the local treatment because at the least they inhibit the filling of the defect by rapid development of granulations and may even cause direct damage to the tissues. Only flavazole 1 in 2 000 or in very special cases of heavy proteus infection 1 per cent in carbowax, is used. Once the infection of the sore has been checked by antibiotics or flavazole wet dressings with sterile saline solution weak boric solutions or Dakin's solution are substituted.

In many instances scars of healed sores are liable to break down and this makes surgical repair necessary. However, it must be emphasized that the chances of success of any plastic operation on a paraplegic are doomed as long as the patient's condition is unsatisfactory—in particular, as long as the disturbed protein balance and anaemia are not fully corrected. In such cases the author considers plastic operation contra indicated. As far as rotation flaps are concerned we have adopted Kilner's technique. This method has the great advantage compared with the method of rotation flaps as used by surgeons in the United States of America that the whole scar is completely covered without forming a new scar in the lumbar region as a result of swinging a flap from that area on to the sacrum. Special mention should be made of a method introduced some years ago for the treatment of ischial sinus sores called pseudo tumour technique (Guttmann 1954).

Of great importance is post operative care without which a successful surgical repair of sores is impossible. As the patient will have to lie post operatively partly or entirely in the prone position especially after surgical repair of sacral sores it is necessary to get him accustomed to this position before the operation. A plastic operation like any other major surgical procedure may easily have an adverse effect on the unstable vasomotor control of these patients and pulse and blood pressure need careful attention during the operation and in the first few days after. Blood transfusions are often necessary to counteract prolonged surgical shock in paraplegia. Haematomas have to be removed immediately. Flexion of hips is not allowed for 3 weeks after complete healing of ischial and trochanteric sores then passive flexion is started and gradually increased until the hips are flexed through 90 degrees until flexion produces no tension of the skin. The patient is usually not allowed out of bed until 4 to 5 weeks after operation. Finally no healing of sores whether by conservative or surgical procedures will be successful if the patient is not instructed about the danger of pressure and how to avoid it. In other words the patient has to be made absolutely pressure conscious and this is most important in the prevention of recurrence of sores and thus in the successful rehabilitation and industrial resettlement of paraplegics.

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fertility and potency since the introduction of the intrathecal prostigmine test (Guttmann, 1949). The author has known several paraplegic women who have given birth by caesarean section to normal children, including one patient with a complete lesion below T 6 and another below T 4. A few male paraplegics with clinically complete lesions, but who have retained the erection reflex, have been able to produce children.

Treatment of intractable spasticity

This condition, especially as far as the violent flexor and adductor spasms are concerned, is one of the worst complications of spinal paraplegia. It prevents the patient's rehabilitation and makes his life intolerable. Good progress has been made in recent years in the treatment of this condition as a result of research into its causes. A number of intrinsic and extrinsic factors may act as irritation stimuli to the unrestrained activity of the spinal cord below the level of the complete lesion. Distension of an internal organ in the paralysed part of the body, particularly the bladder, is one of the most important intrinsic factors and a violent initiator of the reflex spasms. It is obvious that the smaller the capacity of the bladder, the smaller the amount of urine necessary to elicit the reflex response of the skeletal muscles and the more frequently these spasms occur. Of equal importance is distension of the rectum and colon. Any intervening infection—in particular, flare up of infection of the urinary tract, toxæmia from pressure sores and lastly, but by no means least of all, anaemia—also lowers the threshold of reflex activity of the isolated cord.

Another factor in the production of reflex spasms is irritation of sensory organs in contracted joints and tendons. The prevention of contractures, as well as continuous treatment once they have developed, by regular passive movements and hydrotherapy, has proved to be most effective in the treatment of spasticity. The author found that alternating passive movements of spastic legs produced by cycling action in a pedal exerciser worked with the arms, have a remarkably relaxing effect on spasticity (Guttmann, 1949).

Another method of overcoming spasticity is the utilization of certain postural reflexes. With the patient in the supine position, the tone of the flexors in hips and knees is increased. In the abdominal and upright position, the tone of the extensors of hips and knees is increased.

However, in a certain number of paraplegics with complete and incomplete lesions, conservative methods alone are not successful. In incomplete lesions with intractable spasticity, peripheral operations, such as elongation of Achilles tendons or resection of obturator nerves, have proved very effective, especially if conservative treatment is continued after operation. In complete lesions of the spinal cord with intractable spasticity, the most effective method has proved to be the intrathecal alcohol block (Guttmann, 1954). It has the great advantage of obviating the need for major and destructive operations, such as laminectomies with posterior or anterior rhizotomies.

Pressure sores

The treatment of pressure sores has been discussed in previous publications (Guttmann, 1945, 1946, 1954, 1955) and it is beyond the scope of this chapter to go into details. However, it may be stressed that the treatment of sores consists of

CHAPTER 13

LOW BACK PAIN

P. H. NEWMAN

WHEN patients with low back pain have been clinically examined and investigated there emerges a large majority in whom this symptom seems to have a musculo-skeletal origin. Only 10 per cent of these will show organic disease in the remainder the symptoms arise from a mechanically unsound lumbar spine.

Pain may be due to strain ligament injury, disc damage, degeneration or prolapse, spondylosis, spondylolisthesis or secondary degenerative joint disease. These are related under the comprehensive title of lumbar insufficiency and its complications.

BIOMECHANICS OF THE LUMBAR SPINE

The function of the vertebral column is twofold: to act as a central pivot of the skeleton as a whole and to protect the spinal cord and cauda equina.

Were it possible to examine *in vivo* a vertebral column stripped down to bone, ligaments and intervertebral disc tissue, it would present as a strong, resilient, yet comparatively supple structure. It is probable that the degree of force necessary to obtain the full range of movement at the various levels of the column would be surprisingly large, especially in the lumbar region, which is an area designed to take violent shock and an extraordinary degree of strain during physical stress.

In a normal column the load is portioned out among the various segments, the amount rationed by the soft tissue elements. In a column which has been damaged this rationing process breaks down and excessive strain will fall on the remaining elements of that deficient segment.

The bones of the lumbar spine are heavily constructed and are well supported by ligaments, muscle groups and fascia. The chief movements in this area are flexion, extension and lateral flexion, rotation being almost absent. In studying the muscles responsible for these movements it is noted that for each there is a group acting directly on the vertebral column and also, with one exception, a group acting indirectly running from the pelvic girdle to the thoracic cage. The latter group has the advantage of mechanical leverage to the extent of the radial measurement of the thorax. Extension is the exception and has no such muscle group to assist by leverage from the thoracic wall. This is a fact of great interest. It would seem that extension is most in need of additional strength. At a glance it appears that the sacrospinalis works at a mechanical disadvantage and that it is alone in its responsibility of extending the lumbar spine, an action which may involve considerable strain.

There are three main factors that prevent excessive strain—that is to say, three mechanisms that enhance the action of the sacrospinalis:

MANAGEMENT OF SPINAL CORD INJURIES

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attention as a functional entity. A study of the mechanics of this region must reveal how important a structure it is and what immense potential strength it supplies.

Gratz (1931) has shown that the tensile strength of fascia lata is 7 000 pounds a square inch and he has also shown that the elasticity of the tissue under tension diminishes after a third of the breaking load is applied. This he describes as the limit of safe stress. Assuming that the lumbar fascia is as strong and has similar properties, it can be expected to take a load of over 2 000 pounds with complete safety provided that the pull is evenly distributed and not too suddenly applied. Besides giving attachment to muscle fibres the lumbar fascia adds strength to the vertebral mass and checks overflexion. The supraspinous ligament running down the tips of the spinous processes is ideally placed to stabilize each individual vertebra and to check the degree of separation between any two adjoining segments.

The range of movement is controlled by the annulus fibrosus the anterior and posterior longitudinal ligaments and not least by the intervertebral joints themselves. The elastic ligamentum flavum also plays an important part as is shown by its hypertrophy at the site of vertebral instability.

Nerve supply of the lumbar spine

There are two sources of sensory nerve supply to the structures of the vertebral column the posterior primary rami and the sinuvertebral nerves. The root emerging from the intervertebral foramen divides into anterior and posterior primary rami. The posterior ramus passes backwards and to the vertebra below through the intertransverse ligament dividing into a medial and lateral branch. The medial branch supplies the intervertebral joint and runs across the lamina to the base of the spinous process. The lateral branch runs into the substance of the sacrospinalis pierces the lumbodorsal fascia and supplies the superficial tissues.

The sinuvertebral nerve first described by Luschka in 1850 is a recurrent branch leaving the root lateral to the posterior root ganglion and re entering the foramen to run back into the spinal canal. Here it passes upwards to supply various structures the posterior longitudinal ligament the periosteum the dura and the blood vessels. These branches are extremely small and difficult to trace and consequently their exact distribution is controversial. Pedersen Blunck and Gardner (1956) have recently carried out a series of dissections in newborn and adult cadavers and serial sections in foetuses. As a result of experiments on the lumbosacral spine structures in cats wherein they noted the effect on blood pressure respiration and muscle tone of stimulating various structures mechanically and electrically they have shown that these nerves contain pain fibres. The rest of the nerve they suggest consists of proprioceptive and sympathetic fibres. They are of the opinion that the sinuvertebral nerve supplies the posterior longitudinal ligament the dura mater periosteum and blood vessels but they can show no distribution to the annulus fibrosus. This agrees with most authorities (Jung and Brunschwig 1932 Wiberg 1949) but Roope (1940) has demonstrated a nerve supply to this structure.

Origin of back pain

The mechanism and origin of back pain remains obscure. The exact distribution

Design of the individual vertebra

A vertebral body is seated on the disc below pivoting in flexion and extension on the nucleus pulposus which in the lumbar spine, lies slightly behind the centre of the annulus fibrosus. To the back of the vertebral body is attached a lever which is long enough to give a ratio of measurement from its tip to the axis of pivot and from there to the front of the vertebral body of $3\frac{1}{2}$ to 1. A muscle thus acting on the tip of this lever as do the multifidus and the interspinalis has a considerable mechanical advantage in extending each individual vertebra. The spinous process in this area is well designed for this purpose. It is strong and well separated from its fellows. Each is enlarged at its tip in a pear shaped manner broader below giving attachment to the tendons and encased in the tough supraspinous ligament.

The lumbar curve

The antero posterior curves are important to the strength of the spine. In full clinical flexion radiographic examination shows that the lumbar curve is seldom obliterated. In overflexion there is a danger of injuring the posterior soft tissue structures. This curve gives the sacrospinalis a bow stringing effect and results in a mechanical advantage for controlling the lumbar spine.

Locking of the lumbar spine

Lifting a heavy weight from a low position would nevertheless still throw excessive strain on a spine if there were not some mechanism of by passing the load to other groups of muscles. In this case the knees and hips are flexed and the extended lumbar spine is stabilized by a mass action of psoas quadratus lumborum and sacrospinalis which has the combined effect of extension and downward compression. The lumbar spine locked in this position, is protected from the strain of lifting. The weight is raised by extending the knees and the hips with the quadratus femoris and the gluteus maximus respectively.

Function of the sacrospinalis

The sacrospinalis clearly has two main functions extension of the spine and counteraction of the flexion force and pull of gravity. Fick (1911) originally suggested that the spinalis muscle relaxed when the spine was in full flexion. Floyd and Silver (1951) proved this experimentally with the aid of surface electrodes connected to an electroencephalograph. The tracing obtained from the electrodes placed over the sacrospinalis showed minimal activity with the subject standing erect. Bending forward produced vigorous contractions in all four leads until the limit was reached when the contractions subsided and remained quiescent while the patient held that position. In this position the spine was resting on its ligaments. When regaining the erect position an initial extension of the hips occurred followed by another vigorous bout of contractions of the muscle. It is interesting to note that the sacrospinalis did not come into action until the spine had been partly raised by the rotation of the pelvis.

The lumbar fascia

The lumbar fascia, a neglected component of our anatomy has attracted little

LUMBAR INSUFFICIENCY

Theoretical interest has often been focused upon the number of lumbar and sacral vertebrae but this has probably no more significance than to shift the site of intersegmental strain around or cephalad.

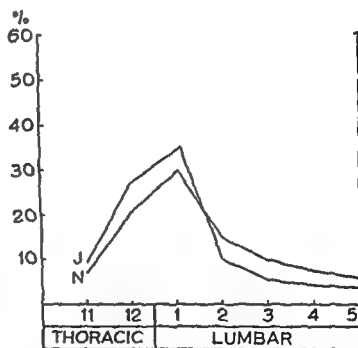
There are two important groups of defects—symmetrical bone development and deficient development of the long vertebral lever.

Post traumatic defect

Essentially the vertebral column consists of the anterior weight bearing and the posterior controlling structures.

The vertebral bodies are injured by violent compression, overflexion, rotation or a combination of these forces. Jefferson (1927) reported 2 006 cases of fracture of the vertebral column and it is interesting to note that in the lower, mobile thoracolumbar area most of the damaged vertebral bodies were at the level of twelfth thoracic or first lumbar. The site of fracture coincides very closely with that given by Nicoll (1949). A graph constructed from these two series of statistics is given—it shows in addition how uncommon are fractures of the lower lumbar vertebrae (Fig. 169).

FIG. 169—Incidence of vertebral fractures at different levels of the thoracolumbar spine in two series of cases. J=Jefferson (1927). N=Nicoll (1949).



The reason for this is not at once apparent—it might be thought that fracture would be most prone to occur where the lumbar spine meets the pelvis. This part, however, is protected by the lumbar curve which is seldom obliterated in a normal spine so that before one vertebral body could impinge on its neighbour severe soft tissue damage would have to occur and this in itself would release the compression force. This is not so at the upper lumbar and lower thoracic level where forced overflexion is liable to damage the vertebral bodies. Nicoll has suggested that lower lumbar body compression occurs when an injuring force is taken with a relatively obliterated lumbar curve—a state that can occur when the legs are braced and the pelvis is tilted backwards. The fulcrum in flexion and extension is the nucleus pulposus. The posterior ligaments may rupture

and function of the two groups of sensory nerves the posterior rami and the sinu vertebral nerves is not clear. It seems that the former distributed to the posterior ligaments tendons fascia and capsules are concerned with the stability and protection of the vertebral column as a whole whereas the latter, supplying the posterior longitudinal ligament dura and periosteum are involved in the mechanism of protection of the nerve tissues

Inman and Saunders (1947) have stressed the clinical differences of the slow dull poorly localized pain arising from mesodermal structures which they term scleratogenous, and the fast, lancinating clearly localized pain of direct nerve stimulation which they term dermatogenous. They believe that pain radiating down the limb can arise from ligamentous tissue. Other workers believe that radiating leg pain typically arises from nerve root irritation or dural sleeve tension and that the ligamentous structures give local pain only.

Kellgren (1939) has produced radiating pain by injecting irritating substances into the posterior ligaments and Hirsch (1948) has obtained the same effect by injecting the disc tissue under tension.

It is not clear how pain arises when a nerve root is involved in the intervertebral foramen by a space occupying lesion. Is it due to pressure on the nerve root to tension on the root or dura or to local chemical change? It is probable that a normal intervertebral disc has no nerve supply. Pain can arise from a damaged disc either from tension against the longitudinal ligaments or from nerve filaments growing into the disc space among granulation tissue.

LUMBAR INSUFFICIENCY

Lumbar insufficiency may be apparent or potential. A latent weakness may not give trouble until exposed by strain injury or awkward and sudden movement when it will start acutely or, after an initial period, a chain of symptoms familiar to the clinician.

The strength of the lumbar spine as elsewhere in the vertebral column is dependent upon the perfection of each individual structure working in consummate action. The bone must be well formed and undamaged the discs intact and turgid the ligaments undamaged the lumbodorsal fascia fully developed and well anchored and the muscles tonic co-ordinated and efficient.

In the lower mobile area that is the lower two thoracic and the five lumbar vertebrae trouble is most prone to occur in its most caudal part especially in the last two intersegmental spaces as here the full strain of the conversion from mobility to immobility is felt. The fifth lumbar vertebra and its anchoring tissues bears most of the brunt of the shock and strain imposed upon it from the superincumbent weight of the trunk. It is unfortunate that in this area congenital defect is most likely to occur.

There are three causes of potential lumbar insufficiency: (1) congenital defect (2) post-traumatic defect and (3) muscle atonia.

Congenital defect

Among the many defects that are encountered it is not easy to select those that are functionally significant. Much statistical evidence has been given one way or the other.

on the back convalescence after a period in bed a strain while lifting childbirth or violent activity in adolescence often however there is no known cause The pain is increased by lumbar flexion for example sitting in a slouched position sleeping on a soft mattress bending stooping or lifting especially with extended knees standing leaning forward over a low object as in washing ironing or cooking

Examination is mostly negative The patient stands with a normal posture or with a slightly increased lumbar curve associated with some spasm of the spinal muscles Active flexion may be slightly limited but like movement in other directions is usually full Straight leg raising may give some pain at its limit by rotating the pelvis but not by nerve root stretching Conduction of the crural equina or of the nerve roots is unimpaired

The patient should be examined in the prone position with two or three pillows placed beneath the pelvis Greatest tenderness is elicited in the midline at the lower lumbar level Some widening between the spinous processes or a deficiency of the supraspinous ligament may be felt There is very little else to find in this type of case but so persistent is the patient that the clinician may be tempted to search for other causes He may associate the trouble with an unstable personality a low morale or general malnutrition The presence of these factors undoubtedly lowers the threshold to pain but the essential clinical syndrome remains as a distinct entity As much harm can be done by informing the patient abruptly that there is nothing wrong as by premature transfer to a psychiatrist or admission to hospital for full spinal investigation The fact that the condition is recognized is a relief to the patient she is assured of her own sensation of pain and encouraged that she is not classed as neurotic

Recurrent lumbago

Lumbago is a vague term; it indicates any acute pain in the small of the back It may be associated with a posture of forward flexion lateral flexion or extension This is an acute pain quite distinct from the chronic nagging ache of lumbar strain and is often of dramatic onset or displays a gradual increase in intensity during 12-24 hours It is this history of dramatic back pain often recurrent that places patients suffering from it in the same category A more detailed study of the clinical findings of each case reveals a great variety of fact The pain may be confined to the back or associated with radiating pain in the buttock thigh or leg Paraesthesiae, numbness weakness or objective signs of root involvement may be present The posture which has been described by Steindler (1955) as the antalgic position varies according to the underlying lesion and is due to a protective muscle spasm The variation of associated clinical findings suggests that the essential pathological lesion is not uniform and the number of different views about the exact cause of lumbago gives support to this suggestion

Contrariwise one attack may differ considerably from another in the same patient Patients with recurrent lumbago may at other times have suffered from sciatica with root signs or the chronic pain of lumbar strain and it would be wrong to assume that these different syndromes do not have some pathological and mechanical connexion

Hult (1954) has taken considerable care to prove the connexion in the pathogenesis of lumbago lumbar insufficiency and sciatica Calculating the incidence

but should they remain intact the disc will inevitably be crushed. Injuries to the neural arch can be caused by hyperextension rotation or a direct blow on the spinous process.

Severe flexion injuries of the spine causing crush fracture or extensive soft tissue injury are comparatively uncommon compared with the multiple injuries and insults to the spine that occur in normal everyday life. The most common is a fall on the buttocks especially if the force is unbroken by the hands for example falling down stairs or falling with the hands laden or in the pockets. As the unfortunate person falls with the legs outstretched in front, the tightened hamstrings rotate the pelvis backwards and the spine falls forwards. There is thus a simultaneous upward compression and flexion force at a time when the protective lumbar curve is obliterated. Excessive or sudden heavy loading of the spine may cause compression injury of a disc.

A latent lumbar insufficiency may be exposed by comparatively minor insults. A forward somersault, a sharp rotation as in golf tennis or squash, a jolt in a vehicle, the lithotomy position under a deep anaesthetic, childbirth or lifting with the knees extended and the lumbar spine flexed are productive of this type of injury.

Muscle atonia

Lumbar insufficiency can be due to excessive heavy work in the presence of chronic atonia caused by general malnutrition and was common in the forced labour camps during World War II.

CLINICAL SYNDROMES

An extensive list of the orthopaedic causes of low backache and the many ways in which the lesions can become manifest would be of little practical value and beyond the scope of this book.

At a clinic designed for the treatment and investigation of low backache 413 new patients reported in one year. Of these 316 presented a clinical syndrome that could be classified in one of four groups: (1) chronic low lumbar strain, 92; (2) recurrent lumbago or sciatica or both, 84; (3) prolapsed intervertebral disc, 85; and (4) degenerative change, 55.

Chronic low lumbar strain

Symptoms of chronic strain occur often in the lower part of the lumbar spine. The syndrome is quite definite though sparse in its symptoms and signs. It is important to note that this syndrome is most likely to be followed, not preceded by other syndromes of lumbar insufficiency, thus it is the first stage of weakness. It has been described in greater detail under the title of Sprung Back (Newman 1952).

The patient is usually in the latter half of the second, third or fourth decade and frequently a female. There is pain across the top of the sacrum, often radiating to both buttocks and the back of the thighs as far as the knees but seldom lower. The pain is described as a dull nagging ache, seldom bad enough to put the patient to bed, but sufficient to produce general fatigue and irritability especially at the end of the day. The onset may be associated with a fall or blow.

SPONDYLOSIS

The posture of the lumbar spine is characteristic. There is obliteration of the lumbar curve and a lateral list either towards or away from the painful side. Movement is markedly restricted in the antero-posterior but not in the lateral plane.

The diagnosis is made more certain and identification of the site of injury more accurate when there is evidence of involvement of conduction of the nerve roots or even of the crura equina.

Degenerative change

Chronic instability leads to degenerative change. The response to instability is a posture of hyperlordosis. It is probable that in most cases ligament instability precedes disc degeneration although in primary injury or metabolic change within the disc the converse may be true.

Generally the chronic pain of low lumbar strain is not associated with diminution in disc space and degenerative joint changes until years later. The following changes which are secondary to chronic instability appear throughout the segmental and intersegmental structures:

- (1) Disc degeneration with diminution of the intervertebral space and intervertebral foramina
- (2) Marginal osteophytosis of the vertebral bodies
- (3) Hypertrophy of the ligamentum flavum
- (4) Osteoarthritic changes in the zygapophyseal joints with subluxation and notching of the laminae by the inferior facets of the vertebra above
- (5) A deficient supraspinous ligament
- (6) Kissing of spinous processes

In the more severe cases of chronic instability these changes are seen in many segments. There is permanent rigid hyperlordosis. The stress and strain of flexion shifts cephalad to the thoracolumbar level where a compensatory kyphosis develops as does a tendency to wedging of the vertebral bodies and disc degeneration.

SPONDYLOLYSIS

Spondylolysis is a radiological diagnosis wherein a break is found in the pars interarticularis in one or both sides of the neural arch without forward subluxation of the part of the vertebra anterior to the lesion. Its clinical manifestation is that of lumbar instability. Symptoms in such cases are generally aggravated by manipulative treatment.

The aetiology of spondylolysis is unproven and certainly one cause is not common to every case but evidence is accumulating to suggest that it is often an acquired defect in the nature of stress fracture.

Stewart (1953) has examined 786 Alaskan skeletons in the National Museum in Washington and has made a study of the incidence of neural arch defect at different ages.

He found that the total percentage of 26.3 is more than four times greater than that found elsewhere. Traditionally the Eskimo works in a stooping position leaning forward without bending his knees and is exposed to falls on rock and ice which is a possible explanation of the high incidence of defect.

of each syndrome in 1 200 workers he found that in those suffering from combined syndromes the incidence of each was significantly higher than that estimated in the whole group indicating the increased liability of a patient suffering from one such syndrome to be afflicted with another. In other words, it seems that there is one fundamental pathology for all these syndromes but that in each there is a difference in detail.

The exciting pathological lesion of an attack of lumbago cannot be diagnosed with certainty. Although the predominant symptom is acute low back pain the associated signs and symptoms may differ considerably.

The typical pain of lumbago has been produced artificially by injecting normal saline solution into the disc substance under considerable pressure (Hirsch 1948). Release of pressure or the addition of Novocain stopped the pain. Hirsch concluded that the pain arose from the nerve cells in the posterior longitudinal ligament and did not suggest that there were nerve endings in the disc substance itself. A similar state of affairs can occur when there is pressure by nuclear tissue on the posterior longitudinal ligament thus shifting damaged annulus fibrosus or cartilage plate. The response to pressure on this ligament is a reflex flattening of the spine. This will have the effect of tightening the posterior longitudinal ligament of increasing the volume of the intervertebral space posterior to the nucleus pulposus and of preventing further prolapse. When the posterior longitudinal ligament is intact the disc substance protrudes to either side but when the ligament is badly damaged central prolapse may occur.

Lumbago in extension or flexion

The response to acute injury of the posterior soft tissue structures is one of extension and this being so it will be noticed that the sacrospinalis stands out prominently on either side of the line of the spinous processes. This is the typical reaction to potential instability of the spine.

On principle the response to a space occupying lesion will be an opening up of the area but to a soft tissue damage and potential instability the response will be a closing down. A hip joint full of pus will abduct and externally rotate but the response to a tear of the Y shaped ligament will be adduction and flexion. Similarly there will be a posture of extension for a loose body in the back of the knee and one of flexion for a tear in the posterior capsule. In the spine the same principles apply. An abnormal body threatening pressure to the cauda equina or nerve roots will be counteracted by flexion but damage of the posterior controlling structures such as the supraspinous ligament the lumbar fascia or the joint capsules or potential instability will stimulate a posture of hyperextension.

Prolapsed intervertebral disc

Among the clinical syndromes associated with low back pain that of prolapsed intervertebral disc predominates as a diagnostic certainty. Typical of the history is some preceding evidence of lumbar insufficiency either the chronic pain of strain or recurrent attacks of mechanical derangement. Onset is often associated with some sudden flexion or rotatory movement or by an awkward or excessive strain. Pain can start suddenly or gradually and can be very severe and lancinating sometimes radiating from the back throughout the length of one lower limb and occasionally both.

instability, hyperlordosis and trauma. The last two occur readily in the toddler.

Facet deficiency

Complete absence of the superior articular processes would theoretically allow forward subluxation of the whole vertebra above without the necessity for modification on its part but in practice it is doubtful whether this ever occurs without alteration of the neural arch.

Partial facet deficiency is more common. A combination of poorly developed facets and soft tissue instability may allow the facets of the vertebra above gradually to grind their way with a glacier like action over or between those below leaving behind grooved remains of what were once articular processes (Fig. 171).



FIG. 171.—Drawing from a case at operation with deficient rough superior sacral facets and a partial sacral spina bifida.

Attenuation

Elongation and narrowing of the pars interarticularis may occur in early life. It is the result of excessive strain on a vertebra which had not at that time completely ossified. The excessive strain arises in combination with bony defect from soft tissue instability, an increased inclination at the lumbosacral angle and from repeated mild traumas.

Lack of continuity

It is an interesting observation that the length of the gap in the neural arch seldom corresponds to the extent of forward slip of the body of the vertebra; it is generally much less. If such a defect had been present since slipping began it would undoubtedly have increased with the amount of slip. The same instability

Periosteal new bone formation and fatigue fractures of the neural arch were common during World War II especially in new recruits undergoing strenuous training. The carrying of heavy packs on their backs had the effect of shifting the line of weight bearing posteriorly thus overloading the neural arches.

SPONDYLOLISTHESIS

The difference between spondylolysis and some types of spondylolisthesis is probably only a matter of degree, dependent upon variations in age of onset, stability and subsequent stresses and strains.

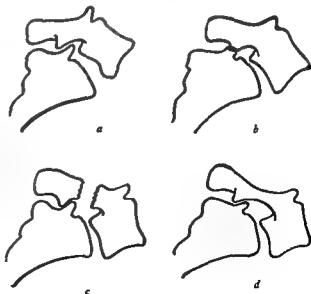
Confusion over the interpretation of the term spondylolisthesis has been partly responsible for the conflicting opinions of incidence and aetiology. The term spondylolisthesis was first coined by Kilian (1853) and was probably designed to include all conditions associated with forward subluxation of the vertebral body, with the possible exception of acute traumatic dislocation or fracture dislocation. Other authorities however have thought a breach in the pars interarticularis to be an essential characteristic for this diagnosis.

Spondylolisthesis can be explained as a manifestation of instability due to bone or soft tissue defect either congenital or acquired. Acute fracture or bone disease is a possible but rare cause of spondylolisthesis. The commonest type of spondylolisthesis probably occurs in infancy or early childhood.

Infantile spondylolisthesis

Spondylolisthesis has never been found before birth, but is common in young children. Considering the design of the vertebra there are three possible theoretical abnormalities that will permit forward slipping: (1) facet deficiency or subluxation, (2) attenuation of the pars interarticularis, and (3) loss of continuity of the pars interarticularis (Fig 170).

FIG 170—This illustration shows several possibilities of bone defect: (a) Normal, (b) Facet deficiency, (c) Break in pars interarticularis, (d) Attenuation of pars interarticularis.



Degrees of facet deficiency occur as developmental abnormalities, but it is probable that all three deficiencies can be acquired in the presence of soft tissue

para interarticularis Adkins (1955) and Gill Munning and White (1955) have reported good results from decompressing the nerve roots by removing the loose neural arch and the surrounding fibrous tissue

Disc prolapse has also been found responsible for nerve root involvement generally arising from the disc above the level of subluxation

Cauda equina lesions

Lesions of the cauda equina have been reported by Arden (1949) Lane (1893) Prip Buus (1943) and Turney (1952) Symptoms may be precipitated by injury in long standing spondylolisthesis In one case they were precipitated by traction on the legs in an attempt to reduce a severe degree of spondylolisthesis Loss of sensation in the buttocks backs of the thighs and calves, and alteration of bladder function are typical findings

TREATMENT

The treatment of low backache of mechanical origin is not at present an exact science it is fraught with obscurity and empiricism Chronic pain arising in the low back can be a severe annoyance a chronic hindrance to normal activities and a handicap to the enjoyment of life Its exact cause is often elusive There is justification therefore for trying various simple forms of treatment always provided that serious disease has first been eliminated and that the treatment is not harmful

A careful clinical and radiological examination should be carried out in all cases In patients with severe incapacitating symptoms it will be necessary to begin some definite treatment at once, but for those with milder pain reassurance and time are possibly the two important elements in treatment

As it would be impossible and of little value to attempt a comprehensive discussion on the vast subject of the treatment of backache two aspects only have been selected as presenting points of interest

Manipulation

Manipulation is a valuable method of treating backache It is used extensively both by the qualified and the unqualified and without doubt is responsible for temporary relief of symptoms in many patients Manipulation is dramatic quick sometimes successful and therefore well suited to certain patients It may be performed with or without a general anaesthetic there is little doubt that the latter method needs a greater degree of confidence and audacity

Whatever manipulation accomplishes it is never more than an unpredictable often irrational and very occasionally a dangerous procedure It is important that the dangers of manipulation be known and the contraindications be recognized by those likely to advise such an undertaking and who intend to maintain an acceptable standard of safety

When a joint is manipulated it is moved within its normal range a part of which may have been lost through spasm or organic change In the spine a rotation movement around both the antero posterior and the longitudinal axis is commonly used Extension longitudinal traction or movements of individual vertebra by pressure on their appendicular processes may be used but flexion

that accounts for the other factors in slip can be responsible for the breach in the neural arch. An attenuated neural arch is liable to fracture while a vertebra in which there is constant stress in the region of the pars interarticularis is prone to fracture. A careful examination of museum specimens in which there is a severe degree of slip will often show a combination of all three elements of defect in the neural arch.

Spondylolisthesis of adult life

There are two other common types of spondylolisthesis. They occur in the adult

Stress fracture in mature bone

Chronic soft tissue instability from ligament insufficiency or disc degeneration throws additional strain on the pars interarticularis and under strenuous conditions stress fracture may occur. The anterior part of the vertebra unsupported by the disc subluxates forwards and sinks down on the vertebra below. The gap in the neural arch opens up wider above than below.

Facet deficiency due to degenerative joint disease

Chronic instability leads in later life to osteoarthritic changes in the zygapophyseal joints. Facet deficiency may occur with forward subluxation of the whole vertebra without neural arch defect. It was originally described by Junghans (1930) as pseudospondylolisthesis, an unnecessary term. Both these types of adult spondylolisthesis are commonest at the joint between the fourth and fifth lumbar vertebrae.

Pain in spondylolisthesis

There are three clinical manifestations in spondylolisthesis: (1) symptoms due to instability; (2) symptoms due to nerve root involvement; and (3) symptoms due to involvement of the cauda equina.

Instability

The most common and typical symptoms of spondylolisthesis are those pertaining to instability. Pain in the back which radiates to both buttocks and the back of the thighs is the usual complaint. This is a pain of mesodermal origin from the ligaments, joint capsules and musculotendinous elements. It can often be relieved temporarily by infiltrating the lumbar fascia, the supraspinous ligaments and the sacrospinalis muscle with local anaesthetic.

Root involvement

Root involvement is not uncommon in spondylolisthesis. It is more likely to occur at the joint between the fourth and fifth lumbar vertebrae than at the lumbosacral joint, particularly in the adult. In the absence of a defect the neural arch is carried forward with the rest of the vertebra, thus encroaching upon the spinal and intervertebral canals. The osteoarthritic changes and thickened ligamentum flavum caused by chronic instability will further narrow these canals.

Some authorities believe that the common pain in spondylolisthesis is due not to stimulation of the afferent nerves in ligaments and capsules but to chronic irritation of the nerve roots by the mass of tissue surrounding the defect in the

pars interarticularis. Adkins (1955) and Gill, Manning and White (1955) have reported good results from decompressing the nerve roots by removing the loose neural arch and the surrounding fibrous tissue.

Disc prolapse has also been found responsible for nerve root involvement, generally arising from the disc above the level of subluxation.

Cauda equina lesions

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TREATMENT

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A careful clinical and radiological examination should be carried out in all cases. In patients with severe incapacitating symptoms it will be necessary to begin some definite treatment at once, but for those with milder pain, reassurance and time are possibly the two important elements in treatment.

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When a joint is manipulated it is moved within its normal range, a part of which may have been lost through spasm or organic change. In the spine a rotation movement around both the antero-posterior and the longitudinal axis is commonly used. Extension, longitudinal traction or movements of individual vertebra by pressure on their appendicular processes may be used, but flexion

never Flexion is a dangerous movement and has probably accounted for some cases of paraplegia

Extravagant claims of good results are made and undoubtedly experience and skill are factors in success but the absence of a properly controlled unbiased follow up may be the cause of overoptimism There are many varied factors in making an accurate estimate of the results of manipulation and this may account for the rarity of published statistics

Coyer and Curwen (1955) have attempted a statistical follow up of 152 patients suffering from backache they were chosen at random They were treated in two ways by manipulation or by bed rest and analgesics with the results shown in the Table

TABLE

<i>Treatment</i>	<i>Well after 1 week per cent</i>	<i>Well after 3 weeks per cent</i>	<i>Persistent symptoms after 12 weeks per cent</i>
Manipulation	50	87	12
Rest in bed	27	60	28

Contra indications

The following conditions contra indicate manipulation (1) absence of a thorough clinical and adequate radiological examination to exclude organic disease (2) old age (3) osteoporosis (4) severe degenerative changes (5) spondylolysis spondylolisthesis and (6) severe pain and spasm or obvious evidence of nerve tissue involvement

Surgical treatment

The treatment of severe back and leg pain of mechanical origin by operation is seldom necessary but in some persistent cases it is the only method likely to give relief This is an immature branch of surgery and the indications for intervention are as yet not always clear but rapid advance has been made during the last decade There are three distinct operative procedures each with different indications none of them is a dangerous undertaking The mortality in four recently published series of low back operations on 618 1250 647 and 350 patients was 2.0 6.0 respectively giving a total mortality of 0.29 per cent It can be assumed therefore that operations in this area performed at well conducted clinics have a mortality somewhere between 0.2 and 0.4 per cent

Removal of disc tissue

Severe prolonged pain which hinders the patient's occupation and does not respond to other means of treatment is the main indication for this operation There should be evidence of root involvement but occasionally operation is indicated for severe pain with back signs only It is a surgical necessity when there is involvement of the cauda equina Careful selection of cases good technique and a controlled course of exercises after operation will give satisfactory results O Connell (1951) published the results of a series of 500 patients operated on for lumbar disc protrusion In 92 per cent there was either complete freedom from symptoms or great improvement

TREATMENT

The modern technique of this operation has evolved by trial and error. Two opposing tendencies, the need for adequate exposure and the desire to harm the stability of the spine as little as possible, have resulted in a routine exposing exposure which can be enlarged should the necessity arise. An interlaminar or partial hemilaminar clearance can easily be extended medially by complete laminectomy or laterally by facetectomy. Unless total laminectomy is necessary it is important to keep intact the spinous processes and supraspinous ligament. These should be sacrificed only in cases of central prolapse of disc tissue or unsuspected more extensive conditions.

A routine operation of this nature should shock the patient very little and necessitate no more than 2 or 3 weeks in bed. Post operative graded spinal exercises starting one week after operation, are an important part of the treatment. As a precaution against root adhesion the sciatic nerve may be stretched during the second week after operation.

Arthrodesis

Arthrodesis is performed for instability and localized severe degenerative change of a disc or the articular surfaces of the zygapophyseal joints. These changes are almost invariably most severe in the lower two lumbar spaces. Before a fusion is advised the severity of the patient's symptoms should be balanced against the long period of immobilization and morbidity and the uncertainty of successful bony fusion.

The time of immobilization advised varies considerably, but most authorities agree that a bone graft in this area is not secure until the end of the fourth month. How much of this period is spent in recumbency and how much in ambulatory immobilization is a matter for individual preference.

Pseudarthrosis rate for lumbosacral fusion

The aggregate percentage pseudarthrosis rate in 932 cases, 430 cases reported by Thompson and Ralston (1949) and 502 cases reported by Cleveland, Bosworth and Thompson (1948) is as follows:

<i>Fusion operation</i>	<i>Number of cases</i>	<i>Percentage pseudarthrosis</i>
5th lumbar vertebra to sacrum	406	6.1
4th lumbar vertebra to sacrum	526	19.4

Method of arthrodesis

Posterior neural arch fusion is the only attested method of arthrodesis. There are three other ways: intercorporeal fusion, both by the anterior and posterior route, and fusion from one transverse process to another. The anterior route has the disadvantage of not revealing the essential pathological lesion and also of having an undesirably high mortality even in competent hands.

Posterior intercorporeal fusion has been advocated by Cloward (1953) who has reported long term cure in 85 per cent of 321 patients. No further large series has been published and judgment on this method must remain in abeyance.

Fusion by placing bone grafts between the transverse processes is still under trial.

Arthrodesis by fusion from one neural arch or spinous process to another has been a recognized method for nearly half a century and has proved to be a valuable surgical procedure. Since the original techniques described by Albee (1911) and Hibbs (1911) many modifications have been devised. Experience gained from 200 lumbosacral fusions using a modification of the method described by Chandler (unpublished) has shown that the following points are important:

- (1) The back of the neural arches and adjoining bone should be carefully cleaned from all soft tissues.
- (2) All ligaments of the segment above the fusion should be left intact for fear of weakening this joint on which much strain will fall in the future.
- (3) The zygapophyseal joints should be excised and packed with bone chips.
- (4) Long grafts cut from the ilium have a natural curve to fit the lumbar concavity and give a maximal area of contact with the sacrum.
- (5) If there is obvious diminution of the intervertebral foramen from abnormal approximation of the vertebrae the neural arches should be wedged apart by an H shaped graft.
- (6) The longitudinal grafts are held in contact with the neural arches by transverse grafts, which can act as H grafts if necessary: wire stretching from the sacral ridge to the spinous process of the highest vertebra involved in the fusion after the method of Chandler, keeps the transverse grafts in place (Fig. 172).

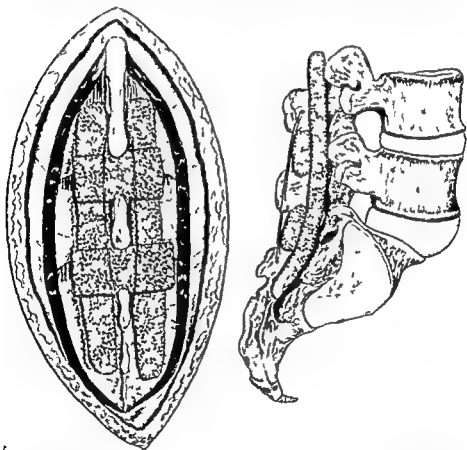


FIG. 172—The method of placing the grafts in Chandler's fusion

- (7) Fresh autogenous bone is preferable to homogenous frozen or boiled bone.
- (8) The present routine for immobilization after operation comprises 3 months recumbency in a plaster bed followed by 6 weeks in a plaster jacket or spinal brace.

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(9) The plaster bed should not be made in hyperlordosis: such a posture will encourage abdominal distension after operation. Fusion in this position will eventually impose great strain on the graft.

Decompression

In a number of cases of spondylolisthesis the cause of symptoms especially radiating pain in the lower limbs is undoubtedly nerve root irritation or tension. In how many is a matter of conjecture but it can be assumed to be the origin of pain in all patients showing signs of impairment of nerve root conduction. In 204 cases of spondylolisthesis Newman (1955) reported a percentage of 14.7. He found nerve root involvement more common with spondylolisthesis at the joint between the fourth and fifth lumbar vertebrae (27.9 per cent) than at the lumbosacral joint (8.5 per cent). For this type of pain a decompression operation was suggested by Briggs and Keats (1947) and later by Adkins (1955) and Gill, Manning and White (1955).

The nerve root is usually compressed at the entrance to or within the intervertebral foramen by the fibrous tissue surrounding the pseudarthrosis of the pars interarticularis or by the margin of the body or the edge of the superior facet of the vertebra below. There is occasionally a disc prolapse usually at a level above the spondylolisthesis.

The operation is designed to relieve the pressure by removing the loose lamina in cases with a neural arch defect. It may also be necessary to remove part or the whole of the superior facet and the projecting margin of the body of the vertebra below.

In spondylolisthesis without a defect in the neural arch in which slipping occurs because of degenerative changes in the joints the root is compressed by the inferior facet of the displaced vertebra and the osteophytosis of the joint. Decompression is achieved by removing the lamina and inferior facets of the intact neural arch.

Decompression is not indicated as a routine procedure because it has three distinct disadvantages. It decreases the soft tissue stability of the two segments, leaving a painful and weak back. It leaves a large gap to be bridged by bone because the lamina, even if loose, acts as a support and source of blood supply to the graft. In addition, removal of the lamina exposes the dura and nerve roots predisposing to adhesion formation.

In summarizing it can be said that decompression is indicated in some cases of spondylolisthesis in which pain is ascribed to root involvement. This is a retrograde step in considering the future strength of the back. Although bone grafting is thus made more difficult it should nevertheless be carried out in all patients who have undergone decompression with the exception of the elderly in whom slipping has occurred with an intact neural arch and severe osteoarthritic changes.

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CHAPTER 14

"FUNCTIONAL" BACKACHE

WILLIAM TIGNER

INTRODUCTION

THE INCIDENCE of psychosomatic illness in Great Britain is still a matter for discussion. Psychiatrists assert that at least one third of those persons who consult their doctors are suffering from functional rather than organic disease. There are others, however, who consider this figure to be too high and believe that a more searching investigation into the pain of which the patient complains will reveal some organic cause. Moreover many patients exhibit a mixture of functional and organic illness and each aspect has its own problem of diagnosis and treatment. In the Rheumatism Centre at the London Hospital where continued re examination and investigation are carried out it has been found that few of the patients are suffering from purely psychosomatic disease. Thus out of nearly 30 000 new patients examined between 1947 and 1955, only 2 per cent were finally found to be suffering from purely non organic disease. On the other hand, Hench and Boland (1946) found that of the men attending the American Forces Rheumatism Centre set up in World War II, no less than fifteen to twenty per cent were suffering from purely psychogenic illness. It is generally agreed that physicians who see large numbers of patients complaining of backache inevitably meet some whose symptoms prove to be largely psychogenic.

AETIOLOGY

The question arises why do some patients who are disturbed in their souls relate their symptoms to their backs? Backache is very common. We are told that the assumption of the upright position by man in his evolution left him with a legacy of potential weakness in his vertebral column. Backache is regarded by some women as an almost inevitable accompaniment of certain periods in their lives. Backache may be due to an almost indefinite number of causes and particularly in the lay mind maladies of many systems outside the vertebral column may be the cause of backache. Men often fear the kidneys are at fault and women the pelvic organs. Moreover it is widely known that the vertebral column houses the spinal cord, and that injuries to this may prove very serious indeed. All these considerations may influence the patient in becoming unduly aware of the back. As backache may be potentially serious it must be taken seriously and the sufferer needs care, sympathy and attention. Thus an inadequate personality may find sympathy and attention if he suffers from backache.

Compensation

In certain patients suffering from backache there arises the question of compensation for injury and ill health. Thus, if a patient after injury to the back does not think he has received or is going to receive adequate compensation for his

suffering he will centre his thoughts on his injured back, and this will continue to agitate him until he obtains what he considers adequate recompense, or, if he does not think the settlement satisfactory he will show himself and the world at large that he is a martyr to his back and to injustice. Similarly, the shadow of National Service, which looms large in the mind of a young man, may, in certain circumstances, cause him to seek refuge in a persistent backache which is obviously incompatible with the life of a soldier.

Hysteria

In most patients there has been at some time in the past an underlying organic cause for backache and this has drawn attention to the back but the backache has been allowed to persist after the cause has ceased to exert its influence. Many reasons can be given for this. In the inadequate personality a backache has revealed a way of escape from the more difficult aspects of life and afforded a chance to shelter at the expense of others. This aspect was well brought out by Halliday (1938). Another person may nurture a grudge against the insurance company who will not pay adequate compensation, or the employer who will not acknowledge liability. Most unfortunately the grudge may be directed against the doctor who has not treated the patient with sympathy and is considered to have been brusque and unkind. It is regrettable but true that some cases of psychosomatic backache derive from medical mismanagement. This may occur in two ways. The matter of a slight backache is passed off lightly as a triviality and the patient far from being reassured is convinced that he is not getting a fair deal but is being neglected. On the other hand backache may be perpetuated and exaggerated by too much investigation and attention. In this respect, an original error in diagnosis may prove disastrous.

An example was found in a young man of 17 years who was referred for examination and opinion by an insurance company in 1955. He was working as a brewer's labourer between leaving school and the call to National Service. A few light crates had slipped from their stacking and had caused a superficial graze of the skin of his back. He was seen by the brewery nurse who applied a gauze dressing gave him some aspirin and sent him back to work but when he told his parents of the episode that evening they immediately took him round to the local hospital. Here unfortunately he was seen by a junior and inexperienced registrar who said the boy had slipped a disc admitted him and applied a plaster jacket. This was a major error for the lad's backache became much worse. He was then referred by his parents privately to a West End consultant who hearing that he had been treated in hospital for a prolapsed disc and had been put in plaster assumed that this diagnosis was correct and admitted him to his own hospital. There all investigations proved negative and a neurologist who examined the boy reported that the only signs present were those of gross hysteria. But the damage had now been done and the parents had instituted legal proceedings against the brewers. The boy was sent to a rehabilitation unit but his stay there failed to relieve his symptoms and he was put on the register of the disabled. Finally he found work as an assembler in a wireless factory where he was treated with consideration as a disabled person. In 1957 the parents not having obtained what they considered a satisfactory offer in compensation for the injury pressed their solicitor for further action. Again the doctors called in by both sides came to the conclusion that the boy had never had a disc lesion. However his hysterical backache is still there and he cannot be parted from a lumbo sacral belt which he wears as evidence of his sufferings.

DIAGNOSIS

In the days before the syndrome of the prolapsed intervertebral disc had been discovered when the diagnoses of 'lumbar fibrositis', 'lumbago' and 'sciatic neuritis' were permissible, the prognosis of these conditions was good and most patients made a satisfactory recovery. With the arrival of the disc lesion, this important fact was at least for the time being forgotten and a wave of enthusiasm for laminectomy as a treatment of choice swept through some schools of surgical thought. Lumbago and sciatica came to be regarded as a surgical disease. In certain hospitals, patients with these diagnoses were lucky to avoid operation. If the diagnosis was correct, there might have been some excuse for this attitude but if the patient's own inadequate personality had drawn attention to the back, the results of laminectomy were sometimes very serious. There was a time after World War II when rehabilitation departments were filled with patients suffering from a failed disc operation. These unfortunates plagued their surgeons who eventually decided that the patients were hysterics of inadequate personality, and that they needed the attention of psychiatrists rather than surgeons. This was probably true, but the diagnosis should have preceded rather than followed operation.

An illustration of this danger is given by the case of a woman who was made of sterner stuff. She was referred for advice in 1949 by her new family doctor, who had just seen her for the first time and the astonishing story came to light. In 1946 she developed backache and was sent at once to a surgeon at a general hospital. He had informed the patient that she was suffering from a disc lesion that the treatment was surgical but that she would have to wait till a bed was available. In the meantime she was put in a plaster jacket. No argument or questioning was permitted and the patient was almost forcibly removed by two members of the nursing staff who applied the plaster-of-paris. The patient did not dare to return to that hospital. It was not till after she had left the district and had found a new doctor that the matter was revived. This was three years later in 1949 and she was still in her plaster jacket! But she was free from backache!

DIAGNOSIS

The diagnosis of psychosomatic backache is fraught with danger. These sufferers are not malingerers. They do experience backache and they do not fall into the traps set for malingerers. This does not make the problem of management or diagnosis any easier. It may seem very obvious to the doctor that the patient has an inadequate personality and is emotionally unstable but it must always be remembered that organic disease can affect such patients as much as those who are more stable. All who are experienced in treating patients suffering from backache will have at some time suspected that a patient is suffering from psychosomatic rather than organic disease only for true organic disease to be revealed later. It is not justifiable to diagnose psychosomatic trouble because examination and investigation reveal no sign of organic disease. This is a refuge for the diagnostically destitute. The diagnosis of psychosomatic backache can only be made if there is good evidence of emotional instability and after meticulous weighing up of the evidence. The diagnosis will depend on the symptoms and signs.

Symptoms

The symptoms while centred on the back will often be very diffuse. The pain often radiates widely, but not according to an anatomical distribution. Thus,

the low backache may spread widely up to the thorax neck and head, and not anatomically down the sciatic nerve. The description given of the pain may be picturesque or even frankly ridiculous. The word 'agony' may frequently be used and yet the patient shows no apparent sign of severe suffering. The periodicity of the pain is often anomalous. Whereas organic backache is often worse after rest, when the muscles are stiff and have to be worked loose, psychosomatic backache is sometimes characterized by its aggravation during the day and by its reaching its peak when the sufferer has to cease all activity and be cared for by others. There are often many other symptoms. Predominant among these may be headache which is sometimes said to radiate from the back over the top of the head. The backache is commonly said to be exhausting and the patient thereby rendered unfit for anything. An inquiry into the general health of the patient may elicit a flood of such symptoms. In spite of exhaustion, patients often complain of inability to sleep. An inquiry into past health reveals a long history of recurrent illness mainly ill defined but incapacitating. Domestic unhappiness and unhappy love affairs may be described, but equally the patient may be reticent about such matters. While some patients are willing if not proud, to call themselves 'highly strung' or 'a bundle of nerves' and may boast of nervous breakdowns, others may resent any suggestion that they are not emotionally stable.

Signs

In clinical examination the patient is often highly demonstrative. The hands are constantly used to point out the various painful areas. The doctor's hand may even be taken and thrust into the area under discussion. These patients find it very hard to relax while under examination and assessment of the range of spinal movement is made very difficult. There is often diffuse tenderness the lightest touch may evoke a withdrawal or even a cry of pain. While these equivocal findings may exist in plenty there is often a dearth of positive signs. Neurological signs do not accord with the symptoms. Lumbar flexion may be present when the patient is asked to sit up on the couch or bed but if the patient is asked to bend forward in the standing position, the flexion is done from the hips while the lumbar hollow is maintained. Radiological and laboratory investigations may be completely normal in spite of the severity of the symptoms.

A case which illustrates many of the points made above is that of a patient who was first seen eight years ago. She was 56 years of age and did some work as a part owner of a milliner's shop. She reported that backache had followed appendectomy ten years before. She had had constant treatment ever since. This had consisted of every known form of physiotherapy a variety of injections and a series of osteopathic manipulations. All these treatments had either failed to relieve the backache or had aggravated it. She said that her general health was poor that she suffered from headaches dyspepsia sleeplessness and fatigue but that she was 'not one to give in'. On examination there was no evidence of organic disease in any system but she was diffusely tender all over the lower back and any movement of the lumbar spine or legs caused wincing and cries of pain. The results of radiological and haematological examination were entirely normal. Any sort of treatment proved quite without effect. physiotherapy aggravated the pain medicines upset her but in spite of this she was a regular and constant attender paying a fee on each occasion for the privilege of talking about her symptoms. She was referred for

orthopaedic and psychiatric opinions she was admitted to hospital for further investigation. No advance of any sort was made. She continues to attend and to explain her illness. She has stopped work as she has enough money of her own for her modest needs. She leads the life of a semi-invalid. She has the sincere sympathy of her friends in her affliction and as she is organically sound she will continue to enjoy ill health for a great many years to come.

MANAGEMENT

The management of psychosomatic backache presents one of the most difficult problems with which a clinician can be faced. The backache is of value to an inadequate personality, and is not likely to be readily discarded. If all investigations lead to the conclusion that the backache is not organic and there are proofs of psychosomatic instability it would seem that the psychological approach is indicated. But here the difficulties start. The patient has a backache which he considers organic and this attitude has almost always been fostered by the preliminary examination and investigation of the first doctor who has seen him and who, quite rightly and properly regarded his pain as organic. Physical treatment has almost always been ordered for a diagnosis postulating an organic cause, perhaps that ready refuge—fibrositis. The failure of such treatment to relieve the symptoms does not worry the patient, who merely asks for something different. Opinions are divided on whether physical treatment should ever be ordered for psychosomatic conditions. One feels very strongly that to order physical treatment for a deranged psyche will only implant in that psyche an impression that the condition is organic. Long experience has proved that these patients do not respond to physiotherapy. Wilson and Tegner (1955) concluded that the prescription of physical treatment was fraught with danger and suggested that it was only justified at the onset of symptoms and for a short time. Physiotherapy particularly in the form of massage can be most pleasant and the patient may find it the only thing that gives relief but the relief is only temporary and massage must be continued indefinitely to the despair of the physiotherapist. More painful and active forms of therapy than massage may be tried but these will aggravate the symptoms and will not be tolerated. Such a situation would suggest that physical treatment is contra-indicated but there is a second trend of opinion which advocates suggestive physical treatment coupled with strong suggestion to the patient that this is going to cure him. The writer has never found this to succeed. Lastly there are those who quite understandably order physical treatment to keep the patient quiet and save themselves the trouble

Psychiatry

It would seem much more reasonable to try patient and painstaking explanation to point out that body and soul are inseparable and that emotional tension can set up muscular tension and perpetuate pain. But the reactions evoked by such an attempt at explanation are varied and none seems satisfactory. The patient may immediately assume that the physician is trying tactfully to explain that there is nothing wrong with him and that he is imagining his pain. This will cause resentment. On the other hand he may assume that the explanation is confirmation that he is seriously ill and needs constant sympathy and attention. The lay term nervous breakdown still has a heroic ring in the lay mind. Many a patient will record with pride that he has suffered a nervous breakdown

while the unfortunate doctor who is trying to manage the case will record yet another hysterical means of escape from an unpleasant environment by an inadequate personality. The most skilled of psychiatrists seem unable to help these people. They are often compelled to report failure, with the comment that suitable guidance in childhood might have prevented these miserable episodes. But the opportunity has been lost.

Drugs

Similarly, drugs do not seem to be of any value in helping these patients. Aspirin is taken in ever increasing quantities and is finally rejected as useless. This in itself is almost of diagnostic significance for most organic pains of the locomotor system respond well to salicylate. The patient frequently runs through all the analgesics and finds that they are either useless or that they upset him. One woman was prescribed aspirin by a synonym by her despairing doctor. Next day she reported to him that the tablets had upset her so seriously that she had to take two aspirins to relieve her symptoms. Nor do the modern anti-depressants, such as Largactil, prove any more efficacious in these patients. The patients all conclude that drugs do not help them.

CONCLUSION

These patients are remarkably successful in achieving their ends through their illness. As a group they receive extraordinary care and comfort from friends and relatives. They are surprisingly selfish at a point which was brought out by Flind and Barber (1945) in their description of psychosomatic rheumatism in the Royal Air Force. The patient with organic backache will be filled with pity and sympathy for the patient with psychosomatic pain.

The conclusion which is generally reached by the despairing doctor is that nothing is going to help these patients for their symptoms are too valuable. I have found no reason to change the opinion I expressed at the Royal Society of Medicine (Tegner 1955) when I said: "What can be done about these patients who in turn exhaust the general practitioners, consultants, psychiatrists and physiotherapists?" The answer is nothing: they will always be with us and the burden must be shared in turn by all the team.

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CHAPTER 15 AN ASSESSMENT OF SPINAL BIOPSY¹

NORMAN L. HICKS POTHAM

VERTEBRAL BIOPSY is an operation to be undertaken with no less reserve than is exploratory laparotomy. It is not an academic exercise but one planned to reveal information that will modify the management of the patient, determine specific treatment and indicate the prognosis. When there is a demonstrable lesion in a vertebra careful thought must be given to the value for proper and intelligent management of having microscopic confirmation of the diagnosis. In spite of all the modern aids to diagnosis the importance of an adequate and careful history and of a complete physical examination cannot be too strongly stressed. The need for special laboratory tests will be apparent after the history taking and examination and when the whole clinical picture is considered together with the radiographic appearances it may be found that a spinal biopsy would add nothing of material value. In certain circumstances the examination of the entire skeleton will indicate other lesions in a site more accessible than the vertebra from where tissue may be taken for microscopic examination.

INDICATIONS AND CONTRA INDICATIONS

When the patient's health or even life depends on an exact diagnosis which can only be made under the microscope, then the means of obtaining it are justified; otherwise they are not. This attitude is reasonable if it is a patient who is being treated and not statistics that are being amassed. However, there is no doubt of the impressiveness and conclusiveness of an analysis of a series of cases of a specific entity well documented and histologically proven. If this goal can be achieved without hazard or harm to the patient it is then highly desirable.

Due consideration must be given to whether the patient has only a solitary lesion in a vertebra or whether other skeletal lesions are present. If the history, physical examination, laboratory aids and radiographic studies fail to elucidate the nature of a solitary vertebral lesion it may be imperative to do a biopsy. The treatment of an inflammatory lesion will be quite different from that of a lesion due to metastatic cancer, and there are instances in which bone involvement was the first indication that a patient had cancer, the primary site of which might not be disclosed until necropsy. In these circumstances a vertebral biopsy is definitely indicated.

When many bones are involved there are usually adequate grounds for regarding the spinal lesion as identical with those elsewhere, but if the radiographs of multiple areas show quite bizarre or different features suggesting that the patient may have more than one disease, it may be necessary to do a biopsy to establish the true facts. Recently in the author's experience a woman aged 61 years was

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and removal of material under direct vision is required. There is such a wide variation in posture, panniculus and pathology that no set rule is applicable for all cases, but it should be remembered that surgical biopsy is a major procedure and every case must be carefully assessed before subjecting the patient to the operation. Admission to hospital is required and the biopsy is performed in the operating theatre under a general anaesthetic. It is usual for the approach to be posterior or postero lateral unless the biopsy is incidental to a transabdominal or intrathoracic procedure in which exploratory findings point to the desirability of carrying out a vertebral biopsy. The technique follows general surgical principles and is not detailed here. Good haemostasis is desirable and especially when malignant tumour is suspected the wound should be closed in layers without any packing and preferably without drains otherwise if malignant tumour is proven drainage and fungation in an open wound may deter or defer radiotherapy which will probably be indicated. Swab smears and cultures of the area subjected to biopsy should invariably be obtained at the time, for what may appear grossly to be tumour may prove on culture to be inflammatory as once happened to the author when a lesion that had the gross appearance of fibrosarcoma produced *Sporotrichum schenckii* on the cultures.

The surgical approach to lesions of the sacrum, the lumbar vertebrae and even the lower thoracic vertebrae may be embarked upon with relative impunity. However, the upper thoracic and cervical bodies present a more serious problem, and needle biopsy methods would seem more attractive.

ASPIRATION BIOPSY

In many instances an open biopsy can be avoided by employment of the aspiration biopsy technique. This is a relatively simple minor procedure usually done with local analgesia, yields a quick diagnosis when positive and has proved satisfactory in about 80 per cent of the author's cases. The greatest drawback to the method is the lack of trained pathologists to interpret the smears so obtained, but this difficulty is being gradually overcome as more pathologists become familiar with the method. The author has never observed any instance where harm has been inflicted (haemorrhage, seeding in the needle track and so on) and we have not obtained any false positives where a benign tumour has been diagnosed as malignant on aspiration biopsy. Most vertebral lesions are admirably suited for this method. The modification using the Turkel needle (Turkel 1951) is available for the central medullary lesions where the cortex is not thin enough to allow entry of the 18 gauge needle usually employed. A more elaborate kit has been devised and described by Craig (1956) and is highly recommended as suitable for practically all circumstances. He has given a detailed description of the technique, variations of approach and of the hazards.

The simpler equipment employing the usual 18 gauge needle and 20 millilitre Luer syringe (Coley 1949) requires more imagination and planning. The Turkel needle obtains a larger and more acceptable plug of tissue and the Craig technique minimizes the hazards by embodying a more replete armamentarium. All methods are materially aided by radiological guidance. Smears or cultures should be taken from the tissue obtained as recommended in open biopsy. Even with the 18 gauge needle, enough tissue is usually obtained for fixation in formalin and processing into the more satisfactory routine sections. This reduces the burden placed

SUMMARY

upon the pathologist as he can interpret organoid patterns rather than scanty clumps of cells

There are areas that can be approached with a needle that would seem hazardous difficult or impossible by open biopsy methods

COMPLICATIONS

In a partially destroyed vertebral body excision of a large biopsy fragment may predispose to collapse. The spinal cord and its membranes must be avoided unless they are involved in the disease. Veins, small and large, are numerous and uncontrolled haemorrhage can prove troublesome. Wound infection is a very unlikely occurrence after aspiration biopsy, but may complicate open biopsy. Fungation of tumour is not a problem with aspiration biopsy, but must be forestalled in open biopsy by avoidance of wound picking. Seeding of tumour along the aspiration needle track is purely hypothetical.

AS THERAPEUTIC SURGERY

The advantage of open biopsy is seen in those rare instances where removal of sufficient tissue for the biopsy is tantamount to removal of the entire lesion. Thus the nidus of an osteoid osteoma could be completely extirpated in the well planned biopsy. A small exostosis or osteochondroma is just as easily removed wholly as in part and saves the patient from a second operation. Although the situation has not arisen in the author's experience, it is conceivable that an early giant cell tumour of a vertebra might give the opportunity for performing a thorough curettage followed by bone grafting as a curative procedure instead of removing a portion of the tumour by surgery and relying on radiotherapy to complete the treatment.

CONCLUSIONS

Biopsy of a spinal lesion is a procedure not to be undertaken lightly but when necessary for elucidation of diagnosis and as a guide to treatment it can and should be done. Either incisional surgical approach or needle aspiration may be employed and radiological guidance will aid considerably in reaching the site of the lesion.

As Turkel (1951) so aptly stated 'The presumptive diagnosis must often be accepted since the vertebral body is surgically not easily accessible and the surgical approach to the vertebral body is a major procedure. The trephine method of biopsy although not a simple method is much easier than the surgical approach from the patient's, the surgeon's and the anesthesiologist's viewpoint.'

Vertebral biopsy can be done if there is good and substantial reason for it. Aspiration or needle biopsy techniques are favoured because of the distinctly lessened morbidity and if the pathology department concerned is in a position to process and interpret the material so obtained.

SUMMARY

Depending upon the size and location of the vertebral lesion, spinal biopsy may be a formidable procedure and therefore not to be undertaken unless there be good reason. It may be essential to establish a diagnosis not ascertainable otherwise by total evaluation of all facts available including complete physical examination, roentgenographic survey and laboratory tests.

Aspiration biopsy by 18 gauge needle or special modification of the needle technique is preferable, but surgical biopsy may be resorted to where indicated. If done the operative wound should be closed without drains or packing. The accessibility of the vertebral lesion will determine the choice of biopsy method and the prospect of obtaining positive information to aid in determining the appropriate treatment to be rendered.

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INDEX

A

- Abscesses
 - mediastinal drainage 146
 - presacral drainage 146
 - prevertebral drainage 146
 - psoas drainage 146
 - retropharyngeal drainage 146
- Acrocephaly, 52
- Acromegaly 239
 - osteoporosis association with 128
- Actinomycosis
 - biopsy confirmation 288
 - vertebral column of 150
 - aetiology 150 151
 - clinical aspect 151
 - radiographic appearances 151
 - treatment 151
- Adrenal S hormone in Cushing's syndrome, 127
- Alkaline phosphatase
 - activity diminished in hypophosphatemia 54
 - level in Paget's disease 123
- Alkaptonuria 53 54
 - causes 53
 - congenital anomalies of the spine in 29
 - symptoms 53 54
- Anaemia
 - Cooley's porosis in 219
 - sickle-cell 218
- Aneurysmal bone cyst 192 197 202-204
 - age incidence 204
 - macroscopical appearance 204
 - microscopical appearance 204
 - radiological appearance 204
 - treatment 204
- Angioma 192
 - aetiology 202
- Ankylosing spondylitis
 - acute onset 159
 - aetiology 155
 - age of onset 155
 - associated complications 162-164
 - associated conditions 162-164
 - cardiac complications 163
 - cervical spine involvement 159
 - chest involvement 159
 - differential diagnosis 164
 - dyspepsia accompanying 164
 - familial incidence 155 156

Ankylosing spondylitis—continued

- female in 162
- incidence 156
- intercurrent disease 164
- iridocyclitis associated 162 163
- iritis associated 162 163
- localized sclerosis in 222
- nodule formation 159 160
- osteoporosis 127
- pathology 156, 157
- peripheral joint involvement 158
- physical signs 160 161
- presenting symptoms 158
- prognosis 172
- pulmonary tuberculosis accompanying 164
- radiology 164-168
 - hip joints 167 168
 - pelvis 167 168
 - sacro iliac joints 164
 - spinal ligaments 166 167
 - thorax 167
 - vertebral bodies 166
 - synovial joints 166
- sedimentation rate, 158
- sex incidence 155
- sheep cell agglutination test 157
- spondylometry 161 162
- treatment 168-171
 - analgesics 169
 - aspirin 169
 - corticotrophin 169
 - cortisone 169
 - deep radiotherapy 170
 - comparative results 171
 - procedure 170
 - side-effects 171
 - hydrocortisone 169
 - phenylbutazone 169
 - physiotherapy 169
 - prednisolone 169
 - prednisone 169
 - surgery 171
- Ankylosis 223 224
- Annular epiphysis 21
- Annulus fibrosus 6 13 14 16
 - adult lumbar region, in 13
 - function of 14
 - lamination of 14
 - posterior part 15 16

Aspiration biopsy by 18 gauge needle or special modification of the needle technique is preferable but surgical biopsy may be resorted to where indicated. If done, the operative wound should be closed without drains or packing. The accessibility of the vertebral lesion will determine the choice of biopsy method and the prospect of obtaining positive information to aid in determining the appropriate treatment to be rendered.

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Cervical spondylosis—*continued*
 operation treatment by 189
 pathological cause 182 183
 radiographs plain 183
 radiotherapy in 188
 spinal puncture 183
 surgery conclusions 191
 nerve root compression 177-180
 anatomy of nerve root 179
 fibrosis nerve root sleeve 180
 innervation segmental pattern 179
 pain 178-179
 analysis 178
 mode of production 178 179
 signs 178
 symptoms 177 178
 protrusions dorso lateral and foraminal 185-188
 conservative treatment 185
 surgical treatment 186
 facetectomy partial 186
 hemilaminectomy 186
 indications for 186
 results 187
 radiotherapy in 188
 site 176 177
 spinal cord involvement 181-185
 spondylitis 174
 treatment 185-191
 Cervico thoracic scoliosis *See* Scoliosis
 Charcot Marie Tooth neuropathy scoliosis in 68
 scoliotic pattern 92
 Chondroblastoma benign 192
 Chondroitin sulphate in nucleus pulposus 21
 Chondroma 207
 Chondromyxoid fibroma 192
 Chondrosarcoma 192 197
 Chordoma 192
 age incidence 198
 clinical course 198
 invasive intracranial sphenoid occipital 198
 macroscopic appearances 199 200
 microscopic appearances 201
 notochordal tissue resemblance to 201
 origin 198
 sacrococcygeal 198
 sex incidence 198
 site 198
 treatment 201
Coccidioides immitis in blastomycosis of vertebral column 152
 Congenital scoliosis correction and fusion 99
 paraplegia in 29
 treatment conservative, 95

Connective tissue congenital deficiency abnormalities due to 50-53
 Crutchfield tongs head traction with 249
 Curvatures of the spine 1-3
 Cushing's syndrome 212
 adrenalectomy in 134
 adrenal S hormone excessive production 127
 fractures associated with 131
 osteoporosis due to 127 128
 distribution in 128
 with vertebral collapse in 218
 rib fractures multiple 218

D

Dermatomyotome 4
 Diaphyseal long bone formation 8
 Diastematomyelia 29 30 48-50
 symptoms 50
 treatment 50
 Diplomyelia *See* Diastematomyelia
 Disc changes 212 215
 Disseminated sclerosis mistaken diagnosis 30

E

Echondrosis physaliphora 198
 Electroconvulsive therapy vertebral colapses following 216
 Eosinophilic granuloma 288
 Epiphyseal ring fusion to diaphysis 10
 Epiphysitis 113
 Ewing's tumour 288
 Exostoses cartilage-capped 207

F

Feet impairment of sensation in spina bifida 48
 Fibromas 207
 Fibrosarcoma 192 197
 Foetus malformation causes
 hormones 30
 hyper vitaminosis 30
 hypo vitaminosis 30
 insulin 30 31
 rubella 30
 x irradiation 30
 Fracture spinal first aid 247 248
 movement of patient 247
 Fragilitas ossium 51 126 217 218
 Friedreich's ataxia scoliosis in 68
 scoliotic pattern 92
 Fungus infections of vertebral column 150-152
 Fusion of the atlas to the occiput, 31-37

- Antero posterior curves
in adult 1 2
newborn child in 1
- Arnold Chiari deformity 30
definition 35
- Arthrodesis low back pain for 277
method 277-279
- Arthrogryphosis multiplex congenita myo-
pathic scoliosis in 92
- Atlanto axial synovial joints range of
movement 26
- Atlanto occipital synovial joints range of
movement 26
- Atlas
derivation 31
fusion associated with Arnold Chiari
deformity 25 36
symptoms 36
to occiput 31-37
treatment 36
with luxation of atlas on the axis 34
35
platybasia 35 36
symptoms 36
treatment 36
subluxation on axis diagnosis 35
treatment 35
uncomplicated fusion 33 34
- Axial skeleton development 4

B

- Backache functional
aetiology 281-283
compensation 281 282
diagnosis 283-285
hysteria 282 283
example 282
management 285 286
drugs 286
psychiatry 285 286
signs 284 285
symptoms 283 285
- Back pain low
origin 265 266
radiography 231 232
treatment 275-279
manipulation 275 276
contra indications 276
dangers 275
surgical 276-279
arthrodesis 277
method 277-279
decompression 279
disadvantages 279
removal of disc tissue 276
- Bladder
imperfect control in spina bifida 48

- Bladder—*continued*
paralysed management in paraplegia
257 258
- Blastomycosis vertebral column of,
151-152
aetiology 151 152
radiographic appearances 152
treatment 152
- Bone
congenital deficiency abnormalities due
to 50-53
density and structural strength 215 216
tissue primary tumours 197-207
- Brachial neuritis treatment 185
- Brodie's abscess 143
- Brucellar spondylitis 147-148
aetiology 147 148
diagnosis 148
radiographic appearances 148
treatment 148

C

- Calcium content estimation by radio
graphy 211
porosis in 210
sclerosis in 210
- Carcinomatous porosis in 218
- Carpus fusion in oxycephaly 52
- Cartilage congenital deficiency abnor-
malities due to 50-53
- Cartilaginous bone ossification in 6
- Centrum ossification of 8
- Cervical myelopathy *See* cervical
spondylosis
- Cervical sclerotome imperfect segmentation
from occipital sclerotome 33
- Cervical spine scoliosis in Klippel Feil syn-
drome 39
- Cervical spondylitis *See* Cervical spon-
dylitis
- Cervical spondylosis 174-191
aetiology 174
clinical syndromes 177
local symptoms 177
intervertebral discs pathological chan-
ges 175
myelopathy 181-185 188-191
conservative treatment 188
differential diagnosis 183
excision of the protrusion 190
graft operation 190
investigations 183
laminectomy 189
results 189
myelography 184
neurological picture factors con-
cerned 181 182
neurological signs 181

L

- Lead poisoning in young children 222
- Leukaemia
 - osteoporosis due to 133
 - porosis in 218
 - spinal lesions 197
- Ligament 22 24
 - anterior longitudinal 24
 - posterior longitudinal 24
- Ligamenta flava 23
- Ligaments self-differentiation of 14 15
- Lipomas 207
- Longitudinal ligaments 22 24
- Low back pain cause 263
- Lumbago acute 22
 - extension or flexion in 270
 - recurrent 269 270
 - symptoms 269
- Lumbar and thoracic idiopathic scoliosis combined 75
- Lumbar curvature secondary 16
- Lumbar fascia 264 265
 - tensile strength 265
- Lumbar insufficiency 266-268
 - causes 266
 - congenital defect 266 267
 - muscle atonia 268
 - post traumatic defect 267
- Lumbar intervertebral discs
 - foetal 16
 - postnatal 16
- Lumbar movement essential features 27
- Lumbar paralytic scoliosis 84 85
 - muscle paralysis anterior abdominals relationship 91
 - erector spinae relationship 91
 - lateral abdominal flexors relationship 91
 - prognosis 84
- Lumbar scoliosis
 - idiopathic 74
 - muscle paralysis lateral flexors causing 86
- Lumbar spine
 - biomechanics 263-266
 - chief movements 263
 - degenerative change 271
 - individual vertebra design 264
 - locking of 264
 - lumbar curve 264
 - nerve supply 265
 - posterior primary rami 265
 - sacrospinalis function 264
 - sinuvertebral nerves 265
 - strength 266
- Lumbar strain chronic low 268 269
 - age incidence 268
 - examination of patient 269
 - sex incidence 268

Lumbar strain—continued

- symptoms 268
- Lumbar vertebra
 - ovoid area 21
 - sacralization of 43-45
 - unilateral and incomplete 45
 - third fracture-dislocation treatment 249
 - trapezoidal body of fifth 45
- Lumbosacral angle
 - apophyseal facets anomalies of structure 47 48
 - coronal orientation 26
 - fusion in spondylolisthesis 63
 - measurement of 2
 - region congenital deformities of 43-48
 - spine disseminated prostatic carcinoma in 123
- Lymphosarcoma
 - lesions in vertebrae 114
 - spinal lesions 197

M

- Marble bone disease
 - calcium content 215
 - radiographic appearances 221
- Mediastinal abscesses drainage 146
- Meningeal nerve 24
- Meningocele accompanying spina bifida
 - 29 48
 - hidden 30 48
- Menopausal osteoporosis *See* Osteoporosis
- Mesenchyme imperfections in segmentation of 29
- Metabolism congenital disturbances changes due to 53 54
- Metacarpus fusion in oxycephaly 52
- Metastases and vascular supply 229-231
- Metastatic carcinoma 192-195
 - bone metastases percentage 192 193
 - haemopoietic bone marrow involvement 193
 - origin 193
 - prostatic serum acid phosphatase high 207
 - spinal metastases aetiology 193 194
- Metatarsus fusion in oxycephaly 52
- Morquio Brailsford chondro osteodys trophy 51
 - causes 29
- Muscles paralysis in spina bifida 48
- Muscular dystrophy in myopathic scoliosis 92
- Myelography 30 233-235
- Myeloma lesions in vertebrae 114
- Myelomatosis 195-197
 - bone marrow involvement, 195
 - bone involved 196

G

- Gaucher's disease porosis in 218
- Giant cell tumour 192
 - of bone tissue 197

H

- Hamstring muscles action of 2
- Hemivertebrae 29 41 42
 - causes 29
 - Klippel Feil syndrome in 39
 - symptoms 42
 - treatment 42
- Hodgkin's disease
 - spinal lesions 197
 - vertebral lesions 114
- Homogentisic acid 53
- Hydatid disease of spine 152-153
 - clinical picture 152
 - diagnosis 152
 - radiographic appearances 152
 - treatment 153
- Hypophosphatasia 54
 - symptoms 54

I

- Idiopathic hypercalcaemia of infants 221
 - lumbar scoliosis 74
 - radiographic appearances 221
- Intermediate idiopathic scoliosis 76
- Interspinous ligaments 23
- Intertransverse ligaments 23
- Intervertebral discs 13
 - antero lateral prolapse in senile kyphosis 113
 - annular protrusions 107
 - annulus fibrosus 103
 - avascularity 22
 - brown degeneration 104
 - senile kyphosis in 112
 - calcification 104 226-228
 - cervical region in 13
 - components of 21
 - degenerations 102-104
 - degenerative changes 22
 - direct injury 105
 - discography 232 233
 - formation of 5
 - horizontal prolapse 107
 - herniations 104-107
 - lumbar foetal 16
 - postnatal 16
 - region in 13
 - mesenchymal structure 103
 - motion studies 232
 - nerves close relationship 23
 - notochordal cells aggregations in 17

Intervertebral discs—continued

- nucleus pulposus 103
 - escape 105
 - site of 21
- nutrition of 13 21 22
- ossification 104
- pathological changes in cervical spondylitis 175
- primitive 5
- prolapse 270
- protrusion or herniation, 29 104-107
 - onset 270
 - pain 270
 - posture of lumbar spine, 271
- protruded 29
- protrusions 104-107
- radiography of 225
- shape 103
- size 103
- spontaneous herniation 105
- structural complexity 13
- thoracic region in 13
- vacuum phenomena 225 226
- vertical prolapse 105 106
 - in senile kyphosis 113
 - Schmorl's node 105
- Intervertebral foramina 23
- Intervertebral joints synovial 1
- Intraspinous tumours, 207

J

- Joints
 - Luschka of 22
 - synovial intervertebral 1

K

- Klippel Feil syndrome 39-41
 - hemivertebrae in 39
 - scoliosis of cervical spine in 39
 - symptoms 39 41
- Kummell's disease 223
- Kyphoscoliosis 94
 - congenital 94
 - laminectomy incision of dura after 100
 - surgery indications 99 100
 - idiopathic 94 95
- Kyphosis
 - adolescent 113
 - Schmorl's nodes in 105 113
 - senile 112 113
 - antero lateral osteophytes in 113
 - prolapse of intervertebral discs 113
 - brown degeneration of intervertebral discs 112
 - vertical prolapse of intervertebral discs 113

Osteoporosis—continued

- blood examination 133 144
- bone biopsy in 134
- clinical history assessment on 129 130
- congenital abnormalities in 218
- Cushing's disease due to 125 127 128
- distribution in 128
- definition 114 125 126
- diagnosis 129
 - biochemical 132 133
 - idiopathic 128
 - radiological 130 132
- gastrectomy after 126
- general spinal neoplastic infiltration 216
- generalized deformation 211, 212
 - deformity progressive collapse 211
 - diagnosis 211
 - fracture deformation 211
- haemopoietic 218 219
- hormonal 218
- hyperparathyroidism in 218
- immobilization due to 127
- incidence 116
- jaundice in 126
- leukaemia due to 133
- localized radiography limitations of 219
- malnutrition following 126
- menopausal 127 217
 - menopause frequency after 127
- neoplastic 218 219
- oestrogens treatment with 127
- osteogenesis imperfecta in 217
- ovarian maldevelopment association with 128
- pathological features comparative 114-116
- pelvis pathological features in 131
- poliomyelitis severe anterior due to 127
- postmenopausal 125 127 217
 - state due to 125
- pregnancy 128 218
- radiological examination radiation required 219
- radiological picture 215
 - findings 217
- ribs pathological fractures in 131
- senile 125
- serum calcium in 133
 - phosphatase in 133
 - phosphorus in 133
- sex incidence 115 127
- symptomatology 129
- testicular atrophy in 218
 - maldevelopment association with 128
- thyrotoxicosis association with 128
- treatment 134-136
 - adrenalectomy 134
 - calcium intake, 134

Osteoporosis—continued

- mechanical supports 134
 - oestrogen administration 134
 - protein intake 134
 - thyroidectomy 136
 - vitamin D intake 134
 - tuberculous disease of the spine due to 127
 - urine calcium in 133
 - vertebral ACTH associated with 218
 - cortisone associated with 218
 - vertebral fractures in 115
 - vitamin C deficiency due to 126
- Osteosarcoma** 192 197
- Ovarian maldevelopment osteoporosis association with** 128
- Oxycephily** 52

P

- Paget's disease** 188-124
- alkaline phosphatase level 123
 - case classical advanced 121
 - clinical features 121 122
 - microscopical changes 120 121
 - osteoclastic resorption 120
 - radiological changes 123 124
 - sarcoma in 122
 - spinal compression in 122
 - sex incidence 118
 - symptoms 122
 - vertebral fractures in 122
 - vertebral lesions 114 118
- Paralytic equinovarus deformity** 30
- Paralytic scoliosis**
- correction and fusion, 99
 - muscle paralysis in 86-92
 - summary of prognosis 85 86
 - surgery need for 99
 - treatment conservative 95
- Paraplegia**
- bladder and bowels care of 259
 - erected broken vertebrae prevention of collapse 254
 - first aid 247 248
 - immediate mobilization without reduction 257
 - initial management 247 248
 - intractable spasticity treatment 260
 - laminectomy disadvantages 256
 - mortality 245 246
 - nutrition 258
 - paralysed bladder indwelling catheter use of 258
 - intermittent catheterization 258
 - management 257 258
 - pressure sores treatment 260 261
 - progressive angulation prevention 253 254
 - re dislocation prevention 252 253

- Myelomatosis—*continued*
 chemotherapeutic agents effects 197
 complications 197
 incidence 195
 myeloma cells 196
 origin 195
 plasma proteins abnormalities in 195
 porosis 218
 proteinuria in 196
 serum proteins abnormal 207
 skeletal lesions 196
 spine of focal bone destruction 196
 paraplegia in 197
 spleen lesions in 195
 treatment 197
- Myocoele 4
- Myopathic scoliosis 92–94
 arthrogyphosis multiplex congenita in 92
 muscular dystrophy in 92
- Myotome 4
- N
- Nerves intervertebral discs
 close relationship 23
 spinal formation of 24
- Neurofibromatosis 52 53
 correction and fusion 92
 diagnosis 95
 kyphosis associated 92
 prognosis 92
 scoliosis in 69
 surgery indications 99
 thoracic curvature in 92
- Neuropathic scoliosis 92–94
- Notochord 17 millimetre human embryo of 17
 ontogeny in 5
 phylogeny in 5
- Notochordal cells
 aggregations in intervertebral discs 17
 intervertebral cluster 18
 mucoid degeneration 17
 proliferation 17
- Nucleus pulposus 6 103
 birth after 19
 chondroitin sulphate in 21
 development 17–27
 eccentric position in adult lumbar intervertebral discs 15
 foetal life in 18
 hydrophilic properties 22
 notochordal cells in 19
 nutrition by diffusion 22
 site of 21
 turgescence 104
 water content 20
- O
- Occiput fusion to atlas 31–37
- Odontoid process 38–39
 congenital absence 38
 fracture 38
 separate 38
 symptoms 38
 treatment 38
- Ontogeny notochord in 5
- Orthograde position in man 1
- Os odontoideum 38
- Ossification
 cartilaginous bone in 6
 centrum of 8
 vertebral arch of 10–12
- Osteitis deformans *See* Paget's disease
- Osteoarthritis
 central atlanto axial joint of 102
 clinical correlations 102
 definition 101
 neuro-central joints of Luschka of 102
 osteophytes in 107
 posterior apophyseal joints of 102
 small diarthroses of vertebral column of 102
- Osteoblastic lesions serum alkaline phosphatase high 207
- Osteoblastoma benign 192
 age incidence 205
 clinical features 205
 histology 206 207
 radiological appearances 205
 sex incidence 205
 terminology 205 206
 treatment 207
- Osteochondritis 113 228 229
- Osteogenesis imperfecta 126 151
 children in 217
 porosis in 218
- Osteogenic fibroma 205
- Osteoid osteoma 207
 giant 205
- Osteoid tissue replacement 126
 diet adequate 126
 genetic background 126
- Osteomalacia pseudofractures 216
 radiological picture 216 221
 secondary hyperparathyroidism 216
- Osteophytes
 definition 107
 osteoarthritis in 107
- Osteophytosis 107–112
 pathogenesis 108–111
- Osteoporosis 125–136
 acromegaly association with 128
 aetiology 216–219
 age incidence 127
 androgens treatment with 127
 ankylosing spondylitis due to 127
 anorexia nervosa in 126
 biliary obstruction, in, 126

Scoliosis—continued

- neuropathic pattern 68
- paralytic diagnosis 68
- prognosis 80
- prognosis 69
- structural infantile idiopathic 71 72
 - resolving 72 73
 - radiographic features 72
 - recognition 67
- syringomyelia in 68
- thoracic adolescent idiopathic 73
 - sex incidence 70
- juvenile idiopathic incidence 73
 - prognosis 73
- infantile idiopathic prognosis 70 71
 - sex incidence 70
- thoraco lumbar idiopathic prognosis 73
 - symptoms 73
- treatment 95-100
 - conservative 95 96
 - Milwaukee jacket 96
 - surgical 96-100
 - age preferable 97

Sheep cell agglutination test in ankylosing
spondylitis 157

Skeletal cryptococcosis of vertebral
column 152

Skull fusion of sutures in oxycephaly 52

Somites

- origin 4
- sclerotome 4
- series in the embryo 4

Spina bifida 48

- lumbosacral neurological symptoms 48
- meningocele accompanied by 29
 - associated with 48
- occulta 30
- symptoms 48

Spine

- anatomy and development 1-28
- congenital anomalies clinical import
ance 29
 - neurological signs 30
 - segmentation due to imperfect 31-50
 - curvatures of 1-3
 - fractured management 248-257
 - postural reduction 248-251
- hydatid disease *See* Hydatid disease of
spine
- non tuberculous infections infecting or
ganisms 142
- pyogenic osteitis aetiology 142
 - acute clinical course 143
 - differential diagnosis 143
 - mortality 143
 - age incidence 142
 - pathology 142 143
 - subacute aetiology 144
 - diagnosis 144 145

Spine—continued

- radiographic appearances 144 145
- treatment 145 146
- tuberculous bilateral costectomy in 139
- costotransversectomy in 139
- operation in active stage 139 140
- paraplegia 140 141
 - treatment 140 141
- regions affected 137
- symptoms 137
- treatment antibiotic 138
 - conservative 138
 - I N A H 138
 - of complications 140
 - O H I H 138
 - P A S 138
 - splints 138
 - streptomycin 138
 - surgical 139 140
- Spondylitis ankylosing *See* Ankylosing
spondylitis
- Spondylitis brucellar *See* Brucellar
spondylitis
- Spondylitis typhoid *See* Typhoid spon-
dylitis
- Spondylolisthesis 47, 56-64
 - adult life of facet deficiency due to
degenerative joint disease 274
 - stress fracture in mature bone 274
 - causes 29 56 57 274
 - diagnosis 61
 - familial 57
 - infantile 272-274
 - causes attenuation 273
 - facet deficiency 273
 - lack of continuity 273 274
 - instability 274
 - root involvement 274
 - interpretation of term 272
 - lumbosacral fusion in 63
 - pain in 274 275
 - cauda equina lesions 275
 - symptoms 59-61
 - treatment 61-64
 - decompression 279
- Spondylolysis 271 272
 - aetiology 271
 - diagnosis 271
- Spondylometry in ankylosing spondylitis
161
- Spondylosis 111 112
 - definition 112
- Sporotrichosis vertebral column of aetio-
logy 151
 - treatment 151
- Sprengel's deformity in Klippel Feil syn-
drome 41
- Silbamidine in myelomatosis 197

- Paraplegia—*continued*
 sexual problem 259 260
 shock treatment 257
 surgical reduction 252
 efficiency in promoting recovery of
 spinal roots 255
 safety of method 254
 simplicity of method 252
 traumatic external fixation by plaster
 cast 252
 postural reduction technique 248–
 251
 vascular disturbances 259
- Paraplegics
 domestic resettlement 246
 industrial resettlement 246 247
- Parasitic infection 239
- Parathormone injections effects on human
 skeleton 221
- Paravertebral plexus 24
- Paraxial bar 4
- Pars interarticularis 10
- Pelvis size in young children 1
- Perichondrium osteogenetic function 10
- Periosteal bone formation of 8
- Phenylalanine defective metabolism in
 alkaptonuria 53
- Phylogeny notochord in 5
- Platybasia
 cause 35
 definition 35
 fusion of atlas associated with 35 36
- Poliomyelitis
 osteoporosis due to 127
 paralytic scoliosis 86
- Porosis *See* Osteoporosis
- Presacral abscesses drainage 146
- Presacral segments mammals in 1
 man in 1
- Pressure erosions 224 225
- Prevertebral abscesses drainage 146
- Primitive intervertebral disc 5
- Pro atlas 37 38
 clinical significance 38
 symptoms 38
- Psoas abscesses drainage 146
- Psoas major muscles action of 2 3
- Pyogenic infections 236–238
- Pyogenic osteitis of spine *See* Spine
- R
- Radiation hazards 239–242
- Radiation necrosis radiographic appear-
 ances 215
- Radiographs lateral 210
- Radiology and low back pain without
 sciatic radiation 231 232
- Rectum imperfect control in spina bifida
 48
- Retropharyngeal abscesses drainage 146
- Ribs
 anomalies accompanying hemivertebrae
 41
 carcinomatous metastases 123
- Rickets
 calcium content 216
 vitamin D resistant 217
- Rubella in pregnancy 30
- S
- Sacrospinalis function 264
- Sacrum
 inclination of in different adults 1 2
 obliquity of superior surface 45–47
 symptoms 46
 treatment 46 47
- Sarcoma of bone tissue 197
- Scheuermann's disease 8 113
- Schmorl's node 105 106
 adolescent kyphosis in 105
- Sciatica sacralization of lumbar vertebra
 in 45
- Sclerosis
 acquired haemolytic anaemia in 222
 ankylosing spondylitis in 222
 breast carcinoma in 222
 Cooley's anaemia in 222
 generalized 221 222
 localized causes 222 223
 myelosclerotic anaemia in 222
 Paget's disease in 222
 sickle-cell anaemia in 222
 vertebrae solitary sclerosed causes 222
 223
- Sclerotome 4
 caudal part 5
 cranial part 5
 last occipital development of 37
- Sclerotic cells 5
 fissure 5
- Scoliosis
 aetiological classification 67
 cervico thoracic prognosis 70
 Charcot Marie Tooth neuropathy in 68
 congenital 41 77 80
 diagnosis 68
 paraplegia in 29
 prognosis 77–80
 correction and fusion indications 98
 Friedrich's ataxia in 68
 hemivertebrae causing 41
 idiopathic 67
 infantile 55 56
 lumbar 74
 prognosis 69
 summary of prognosis 77
 lumbar idiopathic 74
 neurofibromatosis in 52 69

- Stryker turning frame in spinal fractures 251
- Supraspinous ligaments 23
- Syndactylism in oxycephaly 52
- Synostosis of vertebrae 1
- Synovial joints atlanto axial range of movement 26
- atlanto occipital range of movement 26
- Syphilis 238 239
- vertebral column of 148-150
- aetiology 148-150
- clinical aspect 150
- radiographic appearances 150
- serological appearances 150
- treatment 150
- Syringomyelia
- mistaken diagnosis 30
- scoliosis in 68
- scoliotic pattern in 92

T

- Tarsus fusion in oxycephaly 52
- Testicular maldevelopment osteoporosis association with 128
- Thoracic and lumbar idiopathic scoliosis
- age of onset 75
- combined 75
- degree of curve 75
- Thoracic paralytic scoliosis high 80-82
- degree of curve 82
- muscle paralysis
- abdominal muscles relationship 90
- erector spinae relationship 89
- intercostals relationship 89
- latissimus dorsi relationship 89
- muscle paralysis lateral abdominal flexors relationship 89
- pectoralis major relationship 89
- rhomboids relationship 89
- serratus anterior relationship 89
- trapezius relationship 89
- prognosis 82
- sex incidence 82
- Thoracic scoliosis
- adolescent idiopathic degree of curve 73
- age of onset 70
- infantile idiopathic
- degree of curve 71
- lateral curvature 70
- prognosis 71
- rib rotation 70
- treatment conservative 95
- intercostal paralysis relationship to development 90
- juvenile idiopathic degree of curve 73
- age of onset 73
- Thoracogenic scoliosis 94
- pneumonectomy following 94

- Thoracogenic scoliosis—*continued*
- thoracoplasty following 94
- Thoraco lumbar idiopathic scoliosis
- age of onset 73
- degree of curve 73
- Thoraco lumbar paralytic scoliosis 82-84
- degree of curve 84
- sex incidence 82
- Thromboangitis 143
- Trabeculae
- arrangement of 12
- primary medullary bone of 10
- Trapezoidal body of fifth lumbar vertebra 45
- Thyrotoxicosis osteoporosis association with 128
- Tuberculosis
- biopsy confirmation 288
- radiography in 236
- Tuberculous disease of spine *See* Spine
- Tumours
- haemopoietic tissue of 195
- reticuloendothelial tissue of 195
- Typhoid and brucellosis radiography 238
- Typhoid spondylitis
- aetiology 146
- diagnosis 146 147
- radiographic appearances 147
- treatment 147
- Tyrosine defective metabolism in alkaptonuria 53

U

- Urethane in myelomatosis 197

V

- Vertebra
- assimilation of atlas to occiput causes 29
- bodies of 1
- cervical 3
- abnormalities in Klippel Feil syndrome 39
- coccygeal 3
- congenital wedged 42 43
- development and ossification 6 7
- fifth lumbar in spondylolisthesis 57
- fused causes 29
- internal structure of 12-16
- lumbar 3
- ovoid area 21
- primary 5
- sacralization of 43-45
- primitive formation of 5
- sacral 3
- synostosis in coccyx 1
- in sacrum 1

- Vertebra—*continued*
 thoracic 3
 variations in 3 4
 Vertebral arch ossification of 10-12
 Vertebral arches joints of 22
 Vertebral biopsy
 aid to diagnosis and treatment as 289
 aspiration biopsy 290 291
 complications 291
 indications and contra indications
 287 289
 open biopsy 289 290
 therapeutic surgery as 291
 vertebra involved 289
 Vertebral bodies epiphyses in pronograde
 mammals 10
 myelomatosis collapse in 196
 Vertebral body
 collapse disc changes 212
 radiographic image 210
 Vertebral column
 adult general features 1
 early development 4-12
 Vertebral column—*continued*
 function 263
 innervation 24
 movements 26
 extent and nature 26 27
 Vertebral tumours
 aspiration biopsy 207 208
 biochemical evidence 207
 diagnosis 207 208
 hematological examination 207
 open surgical biopsy 207
 sternal marrow puncture 207
 treatment surgical excision 207
 von Recklinghausen's disease 52 53
- Y
- Yaws spinal lesions in 150
- Z
- Zygapophysis 23